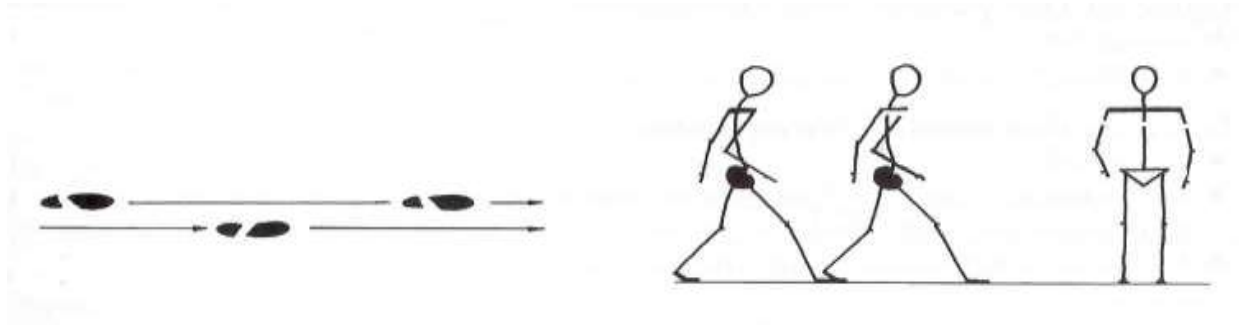


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Pediatric Neurological Examination



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Introduction

The neurological examination of children at various ages can be a challenging feat to any clinician. It is generally felt that examining the neurological system is more difficult in a child than it is in an adult. However, the basic approach to the neurological examination at any age remains the same. The physician begins by determining whether or not a neurological problem exists then proceeds to localizing the involved part of the nervous system in order to formulate a plan of management and treatment. In many instances ancillary tests are needed to confirm the clinical diagnosis. But in spite of the extraordinary paraclinical investigational tools currently at our disposal, the clinical examination remains the most crucial element in this process.

This practical text is written with the intention of helping the students of Al-Quds University medical school and the resident doctors at Makassed Hospital's pediatric department to fully understand the neurological examination of pediatric patients. In its chapters, basic elements of the pediatric neurological examination are detailed. It is hoped that this book contains sufficient practical information to make this examination easier.

To perform the clinical neurological examination properly, some cornerstones are of great importance:

- Always look for symmetry (or asymmetry) between the left and right sides of the body.
- Establishing what is “normal” and making it a reference point to assess the “abnormal” is crucial to reach a precise diagnosis.
- Organize thinking into 8 categories:
 - 1) The “real story” of the present complaint.
 - 2) History (personal and familial).
 - 3) Level of consciousness.
 - 4) Development and behavior.
 - 5) Cranial nerves.
 - 6) Motor functions (posture, movements, tone, power & reflexes).
 - 7) Coordination and gait.
 - 8) Sensory functions.

As you shall see, a lot of figures and illustrations are used to achieve this target. The majority of them are from our patients at Makassed Hospital to whom we feel utmost respect, gratitude and compassion. I hope that this book will help improve the care we provide to them and that in future editions all the figures will be from Makassed.

Lastly, I would like to thank my wife for her support during the two years I spent editing and revising this text. In addition I thank my colleagues at Makassed Hospital's pediatric department: Abdel-Latif, Bassam, Hatem, Imad and Mu'taz, without their assistance this book would not have seen the light.



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Part I

History Taking

1. Age and sex
2. Present complaints (Referral, main complaints, history of present illness and investigations).
3. Prenatal history (pregnancy).
4. Perinatal (delivery) & Neonatal history.
5. Past illnesses (diseases, hospitalizations, surgery, medications,).
7. Family history.
8. Vaccination.
9. Child's psychomotor developmental assessment.

Age & Sex

- Age related diseases (*diseases specific for certain ages*).
- Sex related diseases (*X-linked inheritance, states exclusive for one sex / e.g. Rett syndrome,.....*).

Case presentation

AKMakNP-14551999.....

AGE & SEX

..... A 7-yr. 8-month old male infant was referred to our neuro-pediatric outpatient clinic for evaluation of his abnormal gait that started to deteriorate since the age of 5 yr. Prior to that age, his gait was absolutely normal. Psychomotor development was and still is within the normal range for age.

No significant past medical history.

3 siblings (2 males and 1 female) are clinically well, except one (male), that has an abnormal gait since the age of 4-5 yr.; not investigated. No other positive family history for similar presentation.

The diagnosis of **X-linked** Muscular Dystrophy was evoked on the basis of the following: **Male sex**, wide based gait, important pseudohypertrophy in the calves bilaterally, positive Gowers' sign, decreased muscle power over lower limbs estimated to 4/5, absent knee and ankle DTRs and very high CPK of 19217 IU/L. EMG and NCVs study showed myopathic pattern, excluding the possibility of peripheral neuropathy and confirming the above diagnosis.

Muscle biopsy had confirmed the diagnosis of Muscular Dystrophy. It showed scattered necrotic fibers with degenerative and regenerative changes consistent with muscular dystrophy. Genetic study was not performed due to financial shortage.

*Whether this is **Duchenne** or **Becker Muscular Dystrophy** is to be decided on the basis of ambulation-age. If he continues to walk up to the age of 16-yrs. this will be in favor of Becker MD.*

"This disease is one of the most interesting, and at the same time most sad, of all those with which we have to deal; interesting on account of its peculiar features and mysterious nature; sad on account of our powerlessness to influence its course, except in very slight degree, and on account of the conditions in which it occurs. It is a disease of early life and of early growth. Manifesting itself commonly at the transition from infancy to childhood, it develops with the child's development, grows with his growth - so that every increase in stature means an increase in weakness, and each year takes him a step further on the road to a helpless infirmity, and in most cases to an early and inevitable death."

History of Present Illness

- Chief complaint
Onset in chronologic order.
- If episodic:
Severity, frequency, duration & associated manifestations.
- The exact time of family concern.
- Negative and/or positive Changes *noted since the onset of the illness.*
- Any performed investigations.
- Any diagnostic formulation.
- History of management & therapy.

Case presentation

AOMak1017222005.....

Chief complaint

..... A 6-yr. old male, referred by his pediatrician for neurological evaluation of **sudden onset of limping and pain** that progressed to abnormal unsteady gait (ataxic and wide based). The whole story began one wk. ago and was preceded by URTI.

No recent history of falling down or direct traumas prior to this complaint. No local injections were given recently. No headache or vomiting.

According to the mother, the urinary stream is normal without dripping. Was investigated for Brucella that was negative. CPK, ESR, and ASOT: all were within normal ranges. X-rays of both Rt. and Lt. knees show no abnormalities. CSF showed high protein with no cells. NCVs study was suggestive for myelinic peripheral neuropathy.

Diagnosis: Guillain Barré Syndrome

Case presentation

MAARABC5145242005.....

Episodic details

..... A 3 months old male, evaluated for possible partial seizures, as he started to develop attacks (7-8 times per day) of tonic flexion and adduction of the Rt. upper limb with deviation of the eyes to the Lt. side. Neither shaking, nor changes of his level of consciousness were noted. All started suddenly 3 wks. ago. They took place during sleep and while he was awake.

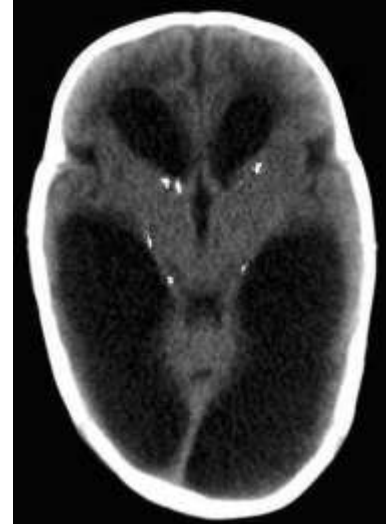
EEG showed epileptic focal activity on the Lt. hemisphere.

4 days ago, he was started on Phenobarbital 3.75mg/kg/day [1 tab of 15 mg q 12 hr.]. With this regimen the attacks became less frequent, brief, and less severe, but of the same pattern.

Prior to the onset of these fits, he was doing well with no history of falling down, direct trauma or fever.

Prenatal History (pregnancy)

- **Gestational age:**
 - Full term (37-42 wk.) ... *completed 37 wk.*
 - Premature (< 37 wk)*causes / if known.*
 - Postmature (> 42 wk)
- **Review of pregnancy**
 - Eventful / Uneventful:
 - Intrauterine presentation
 - Intrauterine fetal movements.
(*present, absent, weak, increased*)
 - Seizures in utero.
(*abnormal movements of fetus*)
 - Bleedings.
 - Oligohydramnios / Polyhydramnios.
 - Intrauterine infections.
(*Mother exposure to viral illnesses, TORCH,...*)
 - Cigarette, Alcohol, Drug use.



Case presentation
RAMak875652004

Intra-uterine infection / Congenital Toxoplasmosis

.....A known x-premature (28 wk) female with **Congenital Toxoplasmosis** (placenta at delivery showed calcifications with cyst formation. Pathological examination showed parasites inside the cyst). The Mother was found to be positive for Toxoplasmosis (IgG and IGM). The patient has high titer of IgG & IgM toxo. In addition she developed Ascitis.

TFU showed large dilated lateral ventricles with normal-sized 3rd and 4th ventricles. Birth weight 1581 gr.

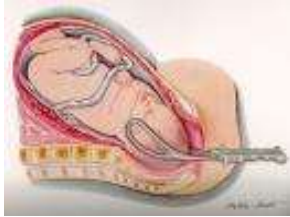
Brain CT showed very significant enlarged lateral ventricles with normal-sized 3rd and 4th ventricle and peri-ventricular calcification.

A diagnosis of obstructive hydrocephalus at the level of Monro foramen was made. In spite of the medication she continues to develop progressive hydrocephaly. IVP shunt was implanted. On the day of operation, HC was 41cm [at 90th percentile]. 20 days later the HC had decreased and became 39.5 cm [at 25th percentile].

Ophthalmic examination showed bilateral Chorioretinal scarring.

Delivery

- Place (*hospital, clinic, home, non-assisted*).
- Normal Spontaneous Vaginal Delivery (*NSVD*).
- Abnormal delivery:
 - Induced labor (*causes*).
 - Cesarean (*causes*).
 - Mechanical instrumentation (*Vacuum, forceps,*



Perinatal & Neonatal History

- General well-being of the newborn.
- Birth weight, length (*not height*), and head circumference (*congenital micro or Macrocephaly*).
- Apgar score.
- Spontaneous breathing (*if cried immediately*).
- Resuscitation (*O2 exposure ; mechanical ventilation*).
- Sucking (*good, weak, absent, gavage feeding*).
- Neonatal jaundice (*mild, severe, phototherapy, blood exchange*).



Signs	Apgar Scoring		
	2	1	0
Heart Rate pulse	Normal >100 beats/minute	<100 beats/minute	Absent (no pulse)
Breathing effort	Normal respiratory effort	Slow or irregular breathing	Absent = apnea (no breathing)
Grimace كشرة To nasal stimulation	Pulls away, sneezes, or coughs with stimulation	Facial movement only (grimace) with stimulation	Absent (no response to stimulation)
Activity muscle tone	Active, spontaneous movement	Arms and legs flexed with little movement	No movement, "floppy" tone
Appearance skin coloration	Normal color all over (hands and feet are pink)	Normal color (but hands and feet are bluish)	Bluish-gray or pale all over

- Score is taken at 1 & 5 minutes after delivery
- Scores obtainable are between 0 and 10 (10 being the highest possible score)

Child's Developmental Assessment

- Very important component of neurological examination.
- **Key point:**
The development of motor control proceeds in a head to toe fashion. The baby first develops head control, then trunk control (sitting), and finally controls the lower extremities (walking). This pattern correlates with the progression of myelination in a head-to-toe direction.
- 4 developmental categories:
 - **Gross Motor**
 - **Fine Motor** (and vision)
 - **Language** (hearing & speech)
 - **Social Interaction**
- It is essential to know the age when any motor, social, and language milestones are normally acquired.



These developmental abnormalities could be:

1. Since birth:

- Since birth, no skills acquisition at all (*absent*).
- Slowing rate of acquisition of new skills.

2. Later in infancy or childhood:

- Arrest of acquisition of new skills (*stagnation*).
- Slowing of the rate of acquisition of new skills after normal rate.
- Arrest of perfection of already acquired skills.
- Regression and /or loss of already acquired skills.

As a result, all the patients ought to be classified as:

- **Normal Development.**
- **Isolated Developmental Delay limited to 1 or 2 or 3 of the 4 categories**
- **Global Delay (involving all the 4 axes)**

- There are several screening tools that can be useful for assessment such as the **Denver Developmental Screening Test II (DDST-II)**.

Gross Motor Development

3-m	Supports Weight on forearms
6-m	Sits momentarily
9-m	Pulls to stand
12-m	Walks with one hand held
18-m	Walks upstairs with assistance (<i>if there is stairs</i>)
24-m	Runs



Fine Motor Development

3-m	Opens hands spontaneously
6-m	Transfers objects
9-m	Pincer grasp
12-m	Releases an object on command
18-m	Feeds from spoon (<i>if allowed or finger feed</i>)
24-m	Builds a tower of 6 blocks (<i>if exposed</i>)



Language Development

3-m	Coos, laughs
6-m	Babbles
9-m	Imitates sounds
12-m	1–2 meaningful words
18-m	At least 6 words (<i>our children are usually delayed in language acquisition</i>)
24-m	2-3 words sentences (<i>very few</i>)

Social Development

3-m	Smiles appropriately
6-m	Shows likes and dislikes
9-m	Plays patty cake
12-m	Comes when called
18-m	Mimics actions of others
24-m	Plays with others

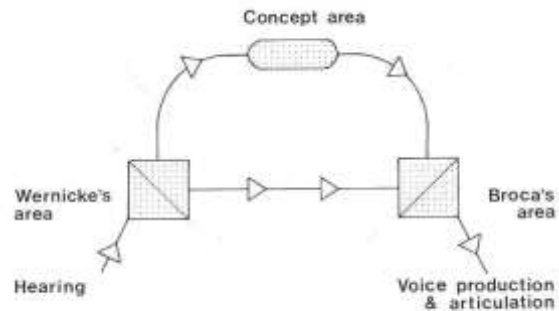


For older children

- School performance
- Behavior
- Physiological autonomy:
 - Eating, dressing, undressing,...
 - Stool & urine Control / full or partial control, no control (primary or secondary encopresis / enuresis).

Normal Speech Development:

1. Starts to cry since the first hours and days of life.
2. Except for crying, the very young baby may make comparatively few sounds besides slight "noises in the throat".
3. By 2 months cooing (يكأغي) a little.
4. By 3 months, chuckles (ضحكات خافتة) may be added to the cooing noises.
5. By 4 months, may laugh aloud.
6. By 6 months babbling (خرخرة المياه) is usually noted; approximately 50% of babies will babble in two or more sounds by this age.
7. By 9 months may say "mama" or "dada".
8. By 1 year add 2-3 words.
9. By 18 months uses about 10 words.
10. By 2 years of age says 2 or 3-word sentences.



Key point:

An infant's ability to use language early is associated with early development of cognitive skills, normal hearing and continuous language stimulation.

- **Dysarthria**
Imperfect articulation of speech due to disturbance of muscular control (think of central / cerebellar or peripheral nervous system damage).
- **Absent/delayed speech** / think of deafness, mental retardation or autism).
- **Lack of intelligibility** / think of mental cerebral palsy)
- **Stammering** (تَهْتِه), **Stuttering** (تَأْتَاه).
To speak with involuntary pauses and repetitions.
- **Dysphonia**
Hoarseness, whispering, high-pitched voice or cry (think of larynx, Myasthenia Gravis, hypothyroidism).
- **Aphonia** / loss of voice (Inability to produce vocal sounds).
- **Vocal tic** of Tourette syndrome.
May include Coprolalia (uttering swear words) or Echolalia (repeating the words or phrases of others).

Denver Developmental Screening Test II (DDST-II)

Objectives:

- To screen children from 1 month to 6 years of age for possible developmental problems in all the 4 axes.
[DDST-II is a screening test, not standardized to determine mental age].
- To gather objective data about suspected developmental problems.
- To monitor children at risk for developmental problems (*ask the parents if they suspect anything*).

Rules:

- Mark the Child's exact age (for FT) and corrected age (for premature) on the score sheet.
- Apply the age line to intersect each functional area.
- Simply determine then whether the child's responses fall into or outside of the normal expected range of success on that item for the child's age.
- Always make sure the child is exposed to the item, such as having stairs at home, rides a tricycle, allowed to spoon or finger feed, before asking the related question.
- This will determine whether the child is classified as:
 - Within the normal range.
 - Within the suspect range.
 - Within the delayed range.

Interpretation:

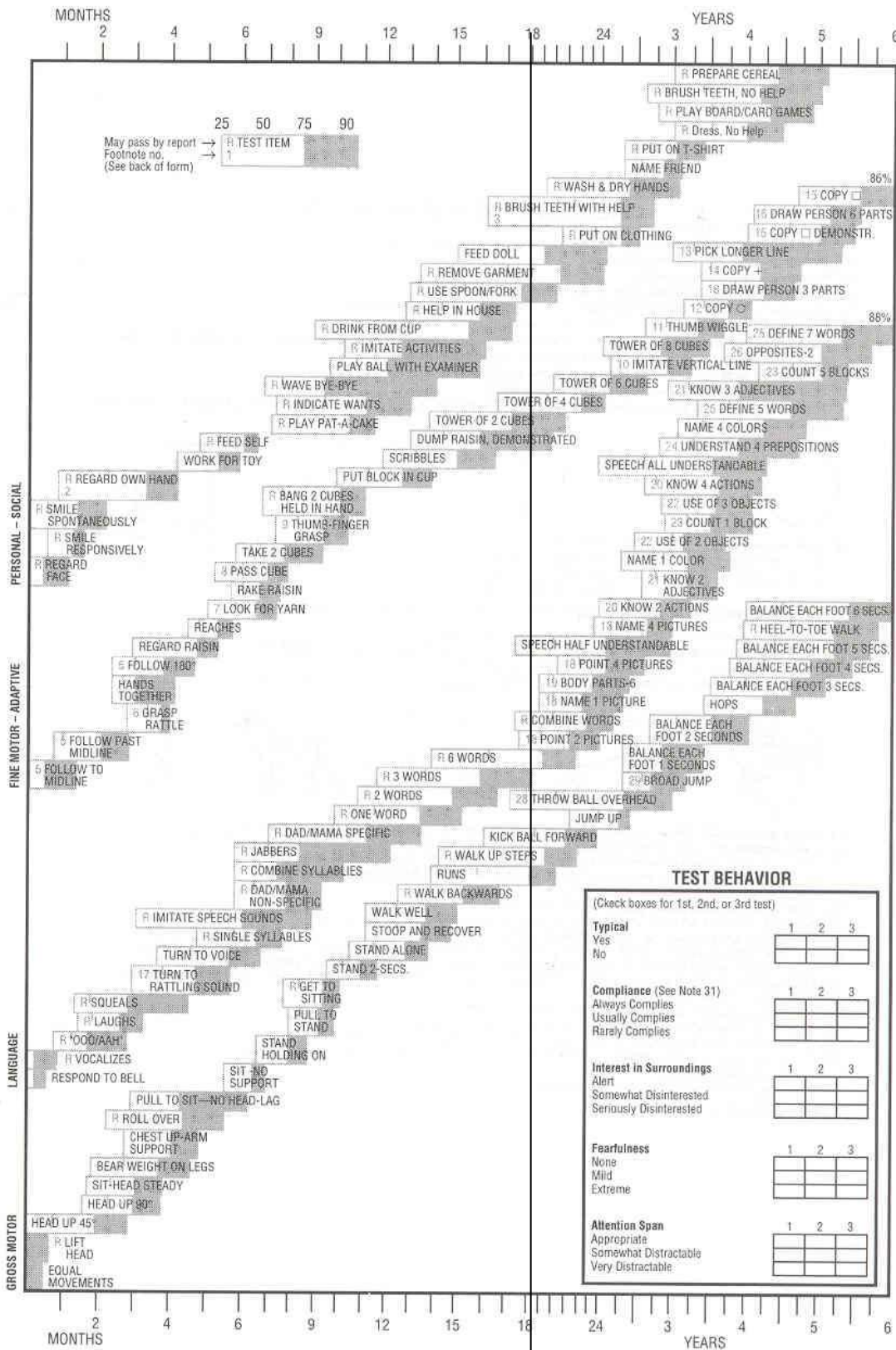
*Category means one of the 4 developmental axes.
Functional area means skills within the same category.*

- **Passes the category** = If passes 3 functional areas within the same category.
- **Caution the category** = If passes 2 + failed 1 functional area within the same category.
Re-test after 1 month, ensure stimulation
- **Fails the category** = if fails 2 or more functional areas.
- **Caution the test** = if fails 1 functional area in more than 1 category.
- **Fails the test** = if fails 2 functional areas in more than 1 category.
Re-test, the whole test, later

Limitations:

No adequate information in the area of language and no differentiation between receptive and expressive language.

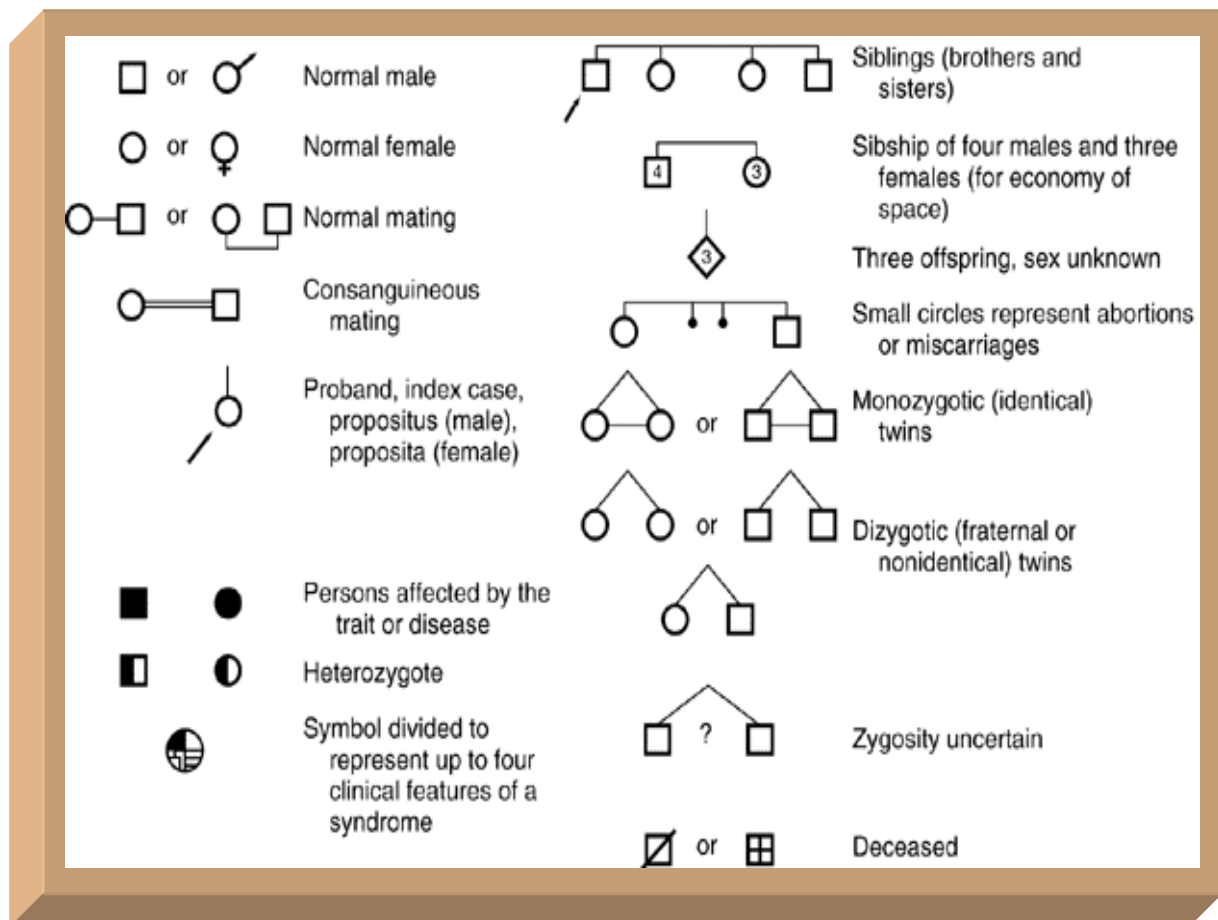
Denver Developmental Screening Test II (DDST-II)



Family History / Pedigree of Family Tree *(extremely important)*

- Parents:
 - Consanguinity
 - Relatives (1st cousins, 1st cousins once removed),
 - Far relatives,
 - Non-relatives.
 - Mother / age / Gravida-Para-Abortions-Still-births (G#P#Ab#Sb#) / state of health.
 - Father / age / state of health.
 - Siblings / # / sort by names / age (DOB) / state of health.
- Similar cases in the family.
- Socio-economic status.

Symbols for constructing a family pedigree:



Part II

Physical Examination

Observation / Inspection



Because the infant and child are unable to fully cooperate for the standard neurological examination, the examination must be tailored to the child and his developmental level and temperament.

The child mostly is examined on the mother's lap.

Observe during:

- Interaction with parents.
- While playing.
- When little attention is directed to the child.



By watching the baby's spontaneous activity, you can collect a lot of information about his mental status, cranial nerves, coordination and motor status.



Leave for the last part of examination, the most threatening and unsettling procedures for the child, such as undressing him, looking at the fundus with an ophthalmoscope, testing the gag reflex or measuring the head circumference.

Overall search for obvious features:

- State of consciousness.
- Behavior during examination (*awareness, hyperactivity of ADHD, Autism*).
- Dysmorphic & characteristic facies.
- Abnormalities of the head (*shape & size*).
- Pattern of cry.
- Unusual postures.
- Asymmetry (*any*).
- Active limb movements (*symmetry, hemiplegia*).
- Abnormal movements.
- Gait (*normal, abnormal*).
- Neural tube defects.
- Joints abnormalities.

State of Consciousness (حالة أو مستوى الوعي)

Generally speaking, being conscious stands for being awake and responsive to one's internal (*such as one's own thoughts, emotions, and bodily sensations*) and external environment. This contrasts with being asleep or being in coma.

Unconsciousness is the lack of reaction to a stimulus. Non/un/in/consciousness exists when consciousness is not present.



It is a synonym of "awareness" and "knowledge" namely our "normal waking life", when we are "fully alert".

The term '**level of consciousness**' expresses how consciousness is described from alert state in one extreme to deep coma in the other one.

Being asleep is the regular state of natural rest observed in all mammals, birds and fish. Sleep is not actually "unconsciousness," but rather, is a natural state of rest characterized by a reduction in voluntary body movement and decreased awareness of the surroundings.

Any state in comparison to fully alert state is termed as "altered states of consciousness". This term describes induced changes in one's mental state, almost always temporary. A synonymous phrase is "altered states of awareness".

State of Consciousness at Birth

- Soon after birth, neonates are alert and ready to interact and nurse. This first alert-awake period may be affected by maternal analgesics and anesthetics or fetal hypoxia.
- Nearsighted مصاب بقصر البصر neonates have a fixed focal length of 20-30 cm, approximately the distance from the breast to the mother's face, as well as an inborn visual preference for faces.
- Hearing is well developed, and infants preferentially turn toward a female voice.
- These innate abilities and predilections increase the likelihood that when a mother gazes at her newborn, the baby will gaze back. This initial period of social interaction, usually lasting about 40 min, is followed by a period of somnolence. After that, briefer periods of alertness or excitation alternate with sleep.

Levels of State of Consciousness

Alert (متنبه - يقظ - حذر)

Is the state of being watchful or ready.

That means the child opens his eyes spontaneously, obeys to verbal command for movements and when spoken to, he is oriented and reacts or converses appropriately.

For the age group 0-23 months, he smiles or coos or speaks appropriately (words or phrases).



Delirium State (هذيان)

Confusion, disorientation, impaired concentration, irritability, agitation, and perhaps excitability.

Lethargic State (كسل - وسن)



Lethargic person awakens with stimulation; drowsy but awake.

Drowsiness is the state of impaired awareness associated with a desire or inclination to sleep and a lack of interest in environment.

Obtundation State (متلبذ الذهن)

An increase in the depth of lethargy. Awake but not alert.

Obtunded person - Mental blunting (متلبذ الذهن); mild to moderate reduction of alertness



Stupor State (انشداد - سبات - خدران - ذهول)

A state of unconsciousness but awakable momentarily with vigorous stimulation. Often precedes coma. Awareness is depressed.

Person in deep sleep; unresponsive but can be awakened with repeated, noxious stimulation. Awareness is depressed but present.

Coma (الغيبوبة – السبات)

Prolonged & profound period of unconsciousness.

Coma differs from sleep in that one cannot be aroused from.



- It involves 2 different concepts:

1) **Reactivity:**

Refers to the innate functions of the brain: the telereceptors (eyes and ears), the nociceptors (responses to pain), the arousal reaction (wakefulness) and the orienting response (turning one's head toward the source of sound or movement). We could also refer to these as **reflexive movements**.

2) **Perceptivity:**

Refers to the responses of the nervous system to stimuli, which have been learned or acquired: language, communication skills, individual methods of movement such as gestures, and less complex learned or acquired reactions such as flinching [اجفال] when threatened. We can also think of these as **conscious movements**.

A person in coma does not exhibit reactivity or perceptivity.

- The person can't be aroused by calling his/her name or in response to pain. Progress of coma is measured by the patient's increasing awareness of external stimuli
- As a person begins to emerge from a coma, they may begin to react to certain stimuli.
- To regain "consciousness" however, reactivity and perceptivity must both be present. These two elements are necessary for a state of awareness.
- The person in coma may exhibit movement, make sounds, and experience agitation. It is important to keep in mind that the comatose patient may exhibit reflex activities which mimic conscious activities.

Glasgow Coma Scale [GCS]

Is the most widely used scoring system for quantifying level of consciousness, mainly, following traumatic brain injury.

It is used primarily because it is (1) simple, (2) has a relatively high degree of reliability and (3) correlates well with outcome.

It is an important tool to assess the degree of brain impairment and to identify the seriousness of injury in relation to outcome.

It involves three determinants:

1. **E**ye opening response
2. **M**otor response
3. **V**erbal response

Glasgow Coma Scale (GCS)

Eye Opening Response	E
Opens eyes spontaneously	4
Opens eyes to verbal command	3
Opens eyes to pain (<i>not applied to face</i>)	2
No eye opening.	1
Motor Response	M
Obeys to verbal command for movements	6
Localizes pain	5
Flexion withdrawal in response to pain	4
Abnormal flexion in response to pain (<i>decorticate posturing</i>)	3
Abnormal extension response in response to pain (<i>decerebrate posturing</i>)	2
No motor response to pain	1
Verbal Response	V
Oriented and converses	5
Disoriented (Confused) and converses (able to answer questions)	4
Inappropriate words	3
Incomprehensible sounds	2
No verbal response	1
Total	15

For children < 5 yr. the verbal response criteria are adjusted as follow

Score	0-23 months	2-5 yr.
5	Smiles or coos appropriately	Appropriate words or phrases
4	Cries and consolable	Inappropriate words
3	Persistent inappropriate crying &/or screaming	Persistent cries and/or screams
2	Grunts or is agitated or restless	Grunts
1	No response	No response

Interpretation

- $E + M + V = 3 \text{ to } 15$ 3 being the worst, and 15 the best.
- **8** is the critical score: ≤ 8 are in coma / ≥ 9 not in coma
- Mild altered consciousness is 13-15
- Moderate altered consciousness is 9-12
- Severe altered consciousness (coma) 3-8

Coma:

No eye opening, doesn't obey commands, no word verbalizations (3-8)

GCS of 11 for example is essentially meaningless, and it is important to break the figure down into its components:

(**E**ye, **M**otor, **V**erbal) such as: **GCS = 11E3M5V3**

Behavior & Attitude during Examination

Normal kids will behave appropriate to their ages, while those for example with attention deficit hyperactivity disorder (ADHD) and with autism will display unusual behavior.

Their behavior during consultation may help in diagnosing them.

Behavior of a Normal Newborn

- Alert and quiet
- Smooth flowing spontaneous movements that are not excessive, jerky or asymmetric.
- Makes attempts to organize and comfort her/himself by sucking on his fists.
- Seems to be attentive to the environment with definite response to bright light directed towards the eyes, consisting of blinking and avoiding the light.
- With repeated stimulus there is habituation (a diminished response to the stimulus).
- Responds to sound by quieting and even turning the head and eyes toward the sound.

Behavior in Autism

A typical **autistic** child's behavior is likely to include:

- Speech disorder (no speech ; delayed development of speech), confusion between the pronouns "I" and "You".
- Lack of interaction.
- Lack of eye contact.
- Stereotyped movements (preoccupation with hands).
- Dislike of being touched.

AUTISM

Persons with autism may possess the following characteristics in various combinations and in varying degrees of severity.



Behavior in ADHD



Those with **ADHD** can't sit still, act before thinking, seem to be "on the go" or constantly in motion. They rush around, touching or playing with whatever is in sight, or talk incessantly. Sitting can be a difficult task. They squirm and fidget in their seats or roam around the room and having difficulty waiting.



Cry

Pattern of cry (intensity & frequency) during the 1st hours and days of life:

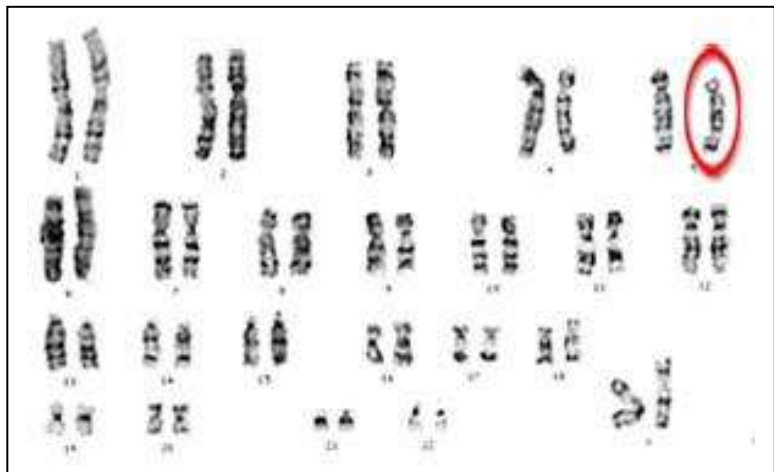
1. High-pitched cry صوت عالي النبرة (*in cerebral irritation or injury*).
2. Hoarse cry (*of congenital hypothyroidism*).
3. Very feeble cry (*of the weak myopathic, SMA, congenital myasthenia*).
4. Peculiar meowing (*cry of Cri du Chat Syndrome*).

Cri du Chat Syndrome



A genetic disorder caused by the loss or misplacement of genetic material from the 5th chromosome.

Affected infants are mentally retarded and have a cat-like cry.



Inspect for Dysmorphic Face

Hypertelorism

Widely spaced eyes by more than the size of one eye due to overdevelopment of the lesser wings of the sphenoid.



Can be found in a variety of syndromes such as craniosynostosis ; Pfeiffer synd ; Fetal immotility sequence (Pena-Shokeir syndrome), chromosomal diseases / trisomy 22



Hypotelorism



Closely placed eyes:

Reduced distance between the eyes, less than the size of one eye.

Causes:

Midline anomalies as septo-optic dysplasia, holoprosencephaly and Scaphocephaly.

Fragile X Syndrome

A chromosomal disease with prominent and elongated ears and long face.



Affected males mostly have mental retardation, and their testes at puberty become larger than normal.



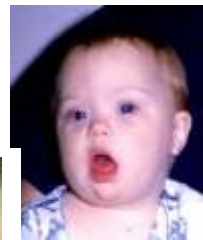
Prevalence: about 1 in 1500 males
Female carriers are typically normal.

Down Syndrome

Trisomy 21

Up-slanting palpebral fissures, Bilateral epicanthal folds, a small nose with flat nasal bridge, open mouth with tongue protrusion, and small ears with over-folded helix.

Karyotype showing trisomy 21 (47,XY,+21)



Coarse Face in Storage Disease

Glutaric Aciduria



Obesity / Over Weight

Pradr-Willi Syndrome



Examination of Ears

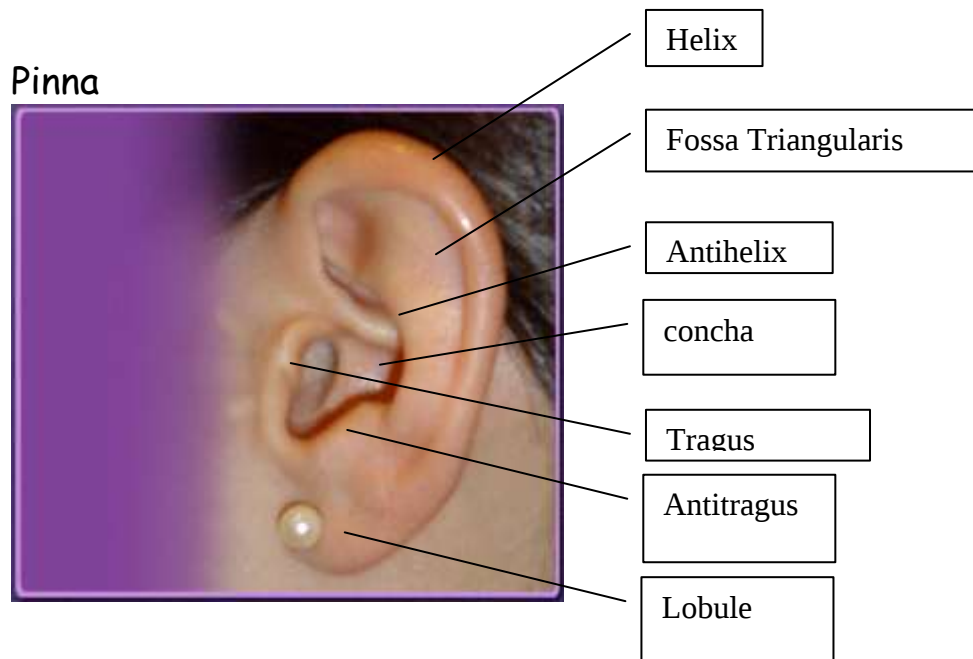
The ears should be inspected for the presence of any deformity, asymmetry, unusually low placement, peculiar slant, cosmetically significant protrusion, skin tags, or other anomalies.

The area immediately in front of the ear should be well examined for the presence of a dermal sinus (avenue for serious infection by staphylococci or other organisms).

A malformed ear may signal the presence of an associated anomaly of the genitourinary tract.

Congenital anomalies of the mouth or nose may also be associated with anomaly in the basic structure of the middle ear or the external ear, with possible deleterious effects on equilibrium and hearing.

Pay attention to signs suggestive of deafness or hearing loss. These are problems that may be due to illness or trauma in the postnatal period, problems that may have been present at birth but overlooked during infancy, and problems in which hearing loss is progressive. Many of these signs relate to speech.



Microtia

Absence of the majority of the pinna صوان الأذن

- Apparent at birth.
- Usually unilateral.
- When bilateral, it may be associated with severe hearing defects.



Low Set Ears



One third of the external auricle should be above an imaginary horizontal line passing through both pupils.



Examination of Hands

All congenital hand malformations should raise suspicion of associated deformities of other organs or tissues.

Hand malformations include **syndactyly** (webbed fingers), **polydactyly** (extra fingers), **Clinodactyly** (crooked fingers معقوف), absent thumb, short fingers and missing fingers.

The unique function of the hand throughout life mandates that function be restored as fully as possible (the ability to pinch and grip is essential to many occupations).



Prominent creases called "flexion creases" appear on the palms of the hands and soles of the feet. People normally have three flexion creases in their palms. Palmar creases develop early, by the 11th to 12th week of life. Abnormalities in palmar creases may indicate problems with early development and be associated with other developmental disorders like Down's Syndrome.

A **Simian** (من قرد) **crease** (جعدة ، ثنية) is a single transverse crease extending across the palm of the hand. It appears in normal people (approximately 1/30). Males are twice as females to have this condition.

*A hand of an infant with Down syndrome.
Note the Simian palmar crease & the shortness
& clinodactyly of the 5th finger.*



Arachnodactyly

Spider fingers

Is a physical condition in which the fingers are long, slender & curved, resembling a spider's legs.



Common Causes: Normal finding, Marfan's syndrome, Homocystinuria

Clinodactyly

Curved finger

Greek "klinein" (to bend, slope or incline) and "dactylos" (finger, toe).



Most frequently curving of the 5th finger (the little finger) toward the 4th finger (the ring finger) called "5th finger clinodactyly".

The basis for the clinodactyly is that the middle bone is underdeveloped and, instead of being rectangular, is wedge-shaped. Clinodactyly is a minor congenital malformation.



It may be an isolated finding in a normal person or in association with congenital malformations and mental retardation.



It is a common component of Down (trisomy 21) and Klinefelter (XXY) syndromes.

Syndactyly

Fused digits or toes



Greek: *syn* means together, and *dactyly* means fingers or digits



Failure of differentiation in which the fingers fail to separate into individual appendages with webbing between fingers or toes.

This separation usually occurs during the 6th-8th wk of embryologic development.



Polydactyly

Extra digit/s

The extra digits may not be immediately apparent unless you count them.



Could be a Congenital genetically inherited abnormality, Autosomal dominant trait.



Some patients may have associated mental handicap and eye abnormalities (*Examine the ocular fundi for any pigmentary retinopathy*).



In some patients, the digits may have been amputated in childhood leaving behind only scar(s).



- Post-axial / in relation with the 5th finger
- Pre-axial / in relation with the 1st finger
- Central / in the middle of fingers.

Fisting Hand



Clenched fist / spasticity of the hand/ spastic flexion of fingers

In upper Motor Neuron Disease

Myopathic Face



- Inexpressive face
- Long thin face
- Restricted eye movements
- Bilateral ptosis
- Drooping mouth (متدلي)
- Inverted V shaped upper lip

Possible causes: All types of Congenital Myopathy

Facial Palsy



- The corner of the mouth on the intact side is pulled toward the opposite side on smiling or crying with inability to close the eye in the affected half.
 - If present at birth, almost certainly, is a peripheral type, therefore affecting the whole of one half of the face. This could be related to birth trauma.
-
- Diff. Diagnosis: peripheral versus central facial palsy.

Paradoxical Breathing

In normal breathing during inspiration, the chest and the abdomen expand outward together in a synchronous way.

In paradoxical breathing, the abdomen expands during inspiration, while the chest indraws (collapses) / discordance of movement.



This is an ominous sign of inter-costal muscular paralysis in Spinal Muscular Atrophy / (SMA type I / Werdnig Hoffmann Disease).

Abnormal Posture (*Body Posture*)

Normal Full Term Neonate

Flexion of all extremities



Tone - Resting Posture at Birth:

For a term newborn the resting posture is flexion of all the extremities with the extremities closely adducted to the trunk.

After the first few days of life, the extremities are still predominantly in the flexed position but they are not as tightly adducted as they are in the first 48 hours of life.

Frog-Leg Posture

Floppy baby in frog-leg posture when lying in supine position



Opisthotonus



The head and the heels are bent backward and the body bowed forward



Kernicterus



Tetanus

Hypertonia in Kernicterus is *extrapyramidal* rather than *cortico-spinal*, affecting the extensor muscle groups with:

- **Retrocollis** (backward arching of neck)
- **Opisthotonus** (backward arching of trunk).

Arthrogryposis

A non progressive condition characterized by multiple (> 1) joint contractures found at birth.

Common causes:

Oligohydramnious, congenital myopathy or neuropathy.



Hemiplegia / Hemiparesis

- Hemiplegia is total paralysis of one side of the body (the arm, leg, and trunk on the same side of the body), whereas hemiparesis is limited to weakness and not paralysis.
- Most common cause: stroke of any origin.
- The earliest sign in hemiplegia is usually absence of movement on one side and a greater tendency to keep the hand clenched than the other.

Todd's Paralysis

Abnormal postures in seizures

- Is a post-ictal focal neurological deficit (paralysis / persistent weakness) following prolonged or a series of seizures.
- The paralysis / paresis resolves spontaneously, usually within minutes or hours, rarely, within days. For that it is often confused with a cerebrovascular event.
- Pathologically, the clinical features may represent neuronal exhaustion after the period of increased activity.

Torticollis

Ipsilateral neck flexion with rotation of the chin to the opposite side.



Ischemic contracture of the Sternocleidomastoid muscle / tumor $\pm$ calcified



Sternocleidomastoid muscle stretches from the sternum to the skull behind the ear

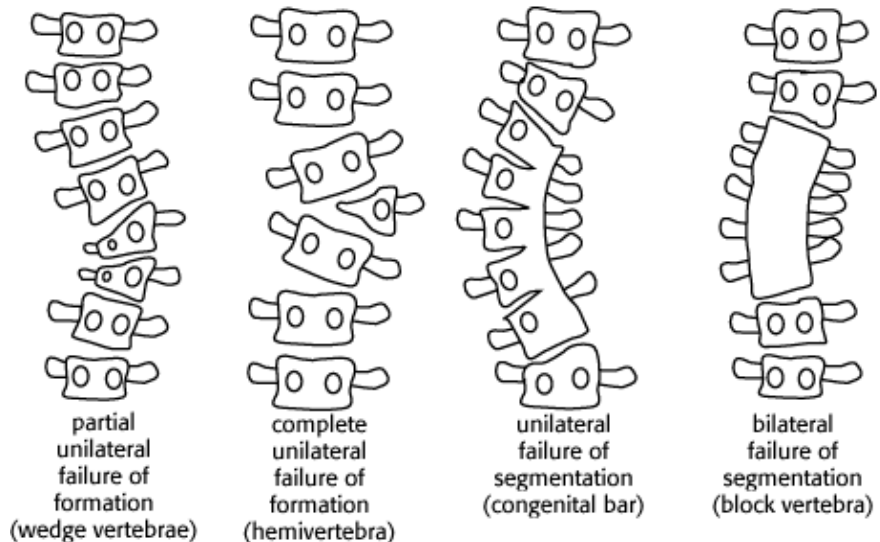
"Wry neck"



Common Causes:

In-utero vascular malformation & birth trauma.

Scoliosis / Curving of the spine in the coronal plane



1. Non-structural or Mobile Scoliosis:

No rotational element.

The curvature is secondary or compensatory to some condition outside the spine. Disappears when the cause is corrected.

eg. leg inequality produces a scoliosis which is corrected by sitting.

2. Structural (True Scoliosis):

The curving is associated with rotation and sometimes, wedging (انفلاق) of the vertebrae.

If associated with pain, consider osteoid osteoma, osteoblastoma or spinal tumor.



Kyphosis



Flexed curvature of the spine in a sagittal plane.

- Is present normally in thoracic and sacral regions (smooth thoracic curvature). Becomes pathological when excessive (exaggerated rounding to the back).
- Most commonly affects the thoracic spine.

1. Postural Kyphosis:

Is common, known as a “round back” or “drooping shoulders”.

May be associated with other defects (flat feet)

2. Structural Kyphosis:

Fixed. Associated with changes in the shape of vertebrae.

Lordosis

(lumbar straightening)

Curving in the spine in a sagittal plane with forward convexity.

A certain amount of lordosis is normal in the lumbar and cervical regions of the spine.

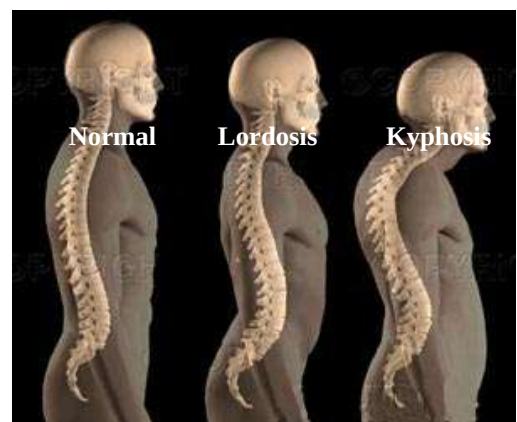
1. Loss of the normal lumbar lordosis:

Sign of paravertebral muscle spasm or other causes.

2. Exaggerated lordosis:

Can occur in:

- Adolescence as a normal variant, pregnancy, obesity
- Vertebral and spinal muscle disease.
- Fixed flexion deformity of the hip.
- Weak muscles of the anterior abdominal wall.



Axial Hypotonia



Infants with generalized hypotonia seem floppy and feel like a "rag doll" *العبة الشرائط* does when held.

Hypotonic infants rest with their elbows and knees loosely extended, while infants with normal tone tend to have flexed elbows and knees.



Head control is mostly absent in the floppy infant, with the head falling to the side, backward, or forward.



To test head lag, pull the child to sitting from lying down.

Note the sitting on the lower end of the back instead of sitting on the buttocks.

Regional Hypotonia / Erb's palsy



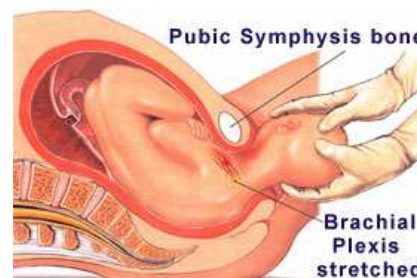
Limp *مترهل* or paralyzed arm and/or lack of muscle control in the arm or hand.

Brachial Plexus injuries can occur at any time, mostly during birth.

During the strain of childbirth, the shoulder of the baby could get caught and stretched behind the Pubic Symphysis bone.

Many babies with Brachial Plexus injuries are larger than average at birth (Macrosomia). However, newborns of all sizes, including premature babies, can have these injuries.

About 1-2 babies in 1,000 suffer Brachial Plexus injuries at birth.



There are 4 types of Brachial Plexus Injuries:

1) **Stretch injuries**-vary depending on the amount of stretching. The nerves will often be compressed from swelling and bruising. Are the least severe and will usually recover within 1 to 2 years with nearly complete function.

2) **Neuroma injuries**-formation of scar tissue that compresses the nerves and may require surgery to restore function.

3) **Rupture injuries**-involve the nerve being torn at several locations and require surgery and therapy to restore normal function.

4) **Avulsion injuries**-when the nerves are pulled from the spinal cord. This is the most severe type of Brachial Plexus injury and requires extensive surgery including a possible muscle transfer to restore function.

The common form is Erb's palsy (affecting the upper part of the brachial plexus, mainly C5, C6), while Klumpke palsy affects the lower parts (C8, T1).

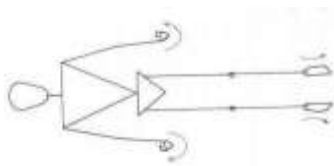
Decerebrate Posturing

Deprived of cerebral function

Characteristic of an individual who has suffered a brain injury that results in neurological dysfunction comparable to that resulting from decerebration of an animal as by having the cerebrum removed, cutting across the brain stem or severing certain arteries in the brain stem.



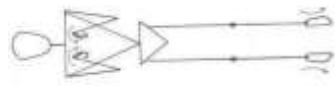
The **decerebrate posturing** is a medical term used to describe the involuntary extending of the upper extremities in response to external stimuli. It is used to measure the severity of a coma.



Decerebrate posturing:

*The shoulders are adducted and internally rotated.
The elbows are extended and the forearms pronated.
The lower extremities are extended*

Decorticate Posturing



Damage to the corticospinal tract, the pathway between the brain and spinal cord.

The upper extremities are flexed at the elbows and wrists, the forearms are pronated, and the shoulders adducted.

The lower extremities are extended.

Decorticate posture may progress to decerebrate posture, or the two may alternate.

- Could be unilateral.

Decorticate posturing is more favorable than decerebrate posturing.

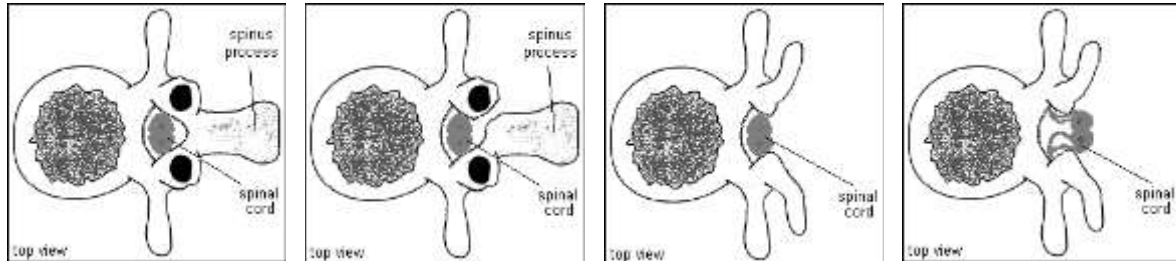
In decorticate posturing, note how the hands are flexed,

while in decerebrate posturing, are extended.

In both the lower limbs are in extension.

Neural Tube Defects

Congenital defects involving the coverings of the nervous system are called neural tube defects.



Normal Vertebrae

Spina Bifida Occulta

Meningocele

Meningomyelocele

Spina Bifida

Latin term: open spine

Spina Bifida Occulta (*Occulta means hidden*)

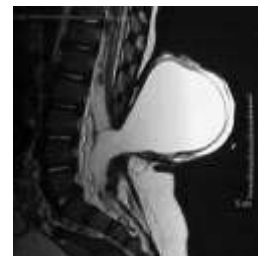
The mildest form in which osseous fusion of one or more vertebral arches is lacking without involvement of the underlying meninges or neural tissue.



A **dimple** somewhere along the spine / often has long, dark hairs sticking out of it. In many cases several vertebra are affected.

Meningocele

A slightly more severe form of spina bifida in which only a sack-like casing filled only with CSF. The spinal cord or brain matter is not herniated.



Spina Bifida Cystica

Myelomeningocele, Encephalomeningocele

The spinal cord and/or brain matter is/are herniated in the sac-like casing.



Examination of the Eyes

Eyes are one of diagnostic indicators of neurological disorders.

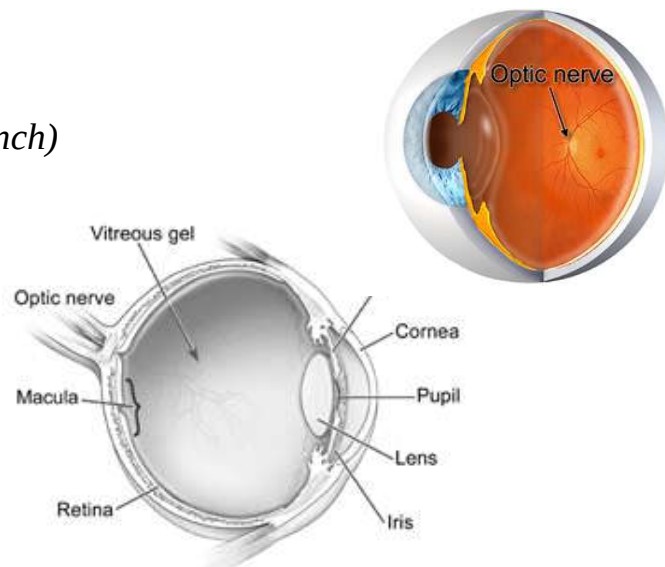
The orbit is the bony cavity that contains the eyeball, associated muscles, nerves, vessels, fat, and lacrimal apparatus.

5 cranial nerves are involved in eyes' movements & functions:

1. Optic nerve
2. Trochlear
3. Oculomotor
4. Abducens
5. Trigeminal (*ophthalmic branch*)

Examination of the eye:

- Inspection
- Pupil reactions
- Ocular movement
- Visual function tests
- Fundus / Ophthalmoscopy



Eyelids Inspection

- The eyeball is guarded by the eyelid, the upper and lower one. The upper is larger and more mobile than the lower.
- The position of the eyelids at rest depends on the tone of the Orbicularis Oculi and Levator Palpebrae Superioris muscles, and on the position of the eyeball.



- Normally the eyelids cover the upper and lower margins of the iris.

Palpebral Fissures

The opening for the eyes between the eyelids



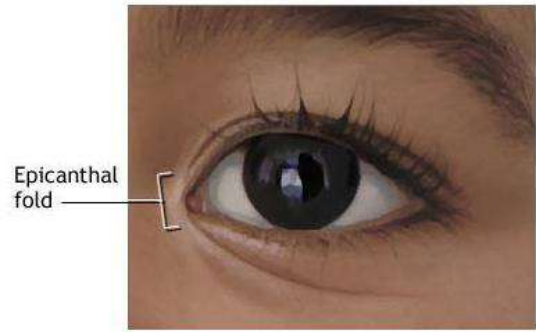
- Should be symmetrical
- They are narrowed in:
 - Blepharospasm (spasm of the eyelids)
 - Photophobia (light sensitivity) associated with migraine and meningeal irritation.
- Upward Slanted in trisomy 21.

Epicanthal Folds

Is the extra skin of the upper eyelid (from the nose to the inner side of the eyebrow) that covers the inner corner (canthus) of an eye.

- Normal in Asians.
- Common in [Down syndrome](#).

May also be seen in young children of any race before the bridge of the nose begins to elevate.



Inability to Close the Eyelid

- Unilateral
- Bilateral

Ptosis

Drooping of eyelids. A person with ptosis is not able to lift one or both upper eyelids to uncover the eye completely.

It could be **unilateral** or **bilateral**

Ask if time-day related as in MG (absent upon awaking from sleep then appears progressively with time and activity).

Causes:

- Levator Palpebrae Superioris (LPS) paralysis, which could be congenital or acquired as in IIIrd nerve (Oculomotor) palsy (uni or bilateral) and Acquired Levator Dehiscence Ptosis where Levator muscle tendon may loosen or detach, causing ptosis.
- Sympathetic paresis (Horner's syndrome)
- Ocular myopathy (usually bilateral drooping).
- Myasthenia Gravis (bilateral day-time related)



*MG before & after
Tensilon test*



Lid Retraction



In hyperthyroidism, usually associated with lid lag and proptosis. Retraction is due to spasm of LPS m. as it is partly innervated by sympathetic fibers.

Capillary Hemangioma



Of the eyelids and one side of the face (port wine stain) as seen in Sturge Weber syndrome. Check for glaucoma, meningeal involvement and epilepsy.

Eyeballs Size

- Equal, non-equal
- Unilateral / Bilateral
- Prominent (Exophthalmos)
as in Thyrotoxicosis
- Sunken (Enophthalmos).
- Enlarged (*congenital or acquired Buphthalmos*).



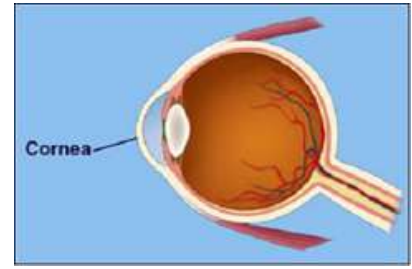
Orbital / Periorbital Cellulitis

	Orbital	Periorbital
Proptosis	+	-
Chemosis (<i>swelling or edema of the conjunctiva</i>)	+	-
Decrease or loss of visual acuity	+	+
Loss of eye movements	+	-
Painful eye movements	+	-



Cornea

Normally transparent with blood vessels only at Limbus (*junction of cornea and conjunctiva*).



Kayser Fleischer Ring



In Wilson Disease due to copper deposition in the periphery of the cornea.

Exposure Keratitis

Dryness as in Facial nerve palsy.



Loss of Corneal Reflex

In Trigeminal nerve palsy which makes the cornea vulnerable to trauma and infection.

- Efferent / Optic n.
- Afferent / Trigeminal n.



Iris

Abnormal Iris Pigmentation



At birth the iris has a slate gray color. It achieves its adult color after about 6 months.

Heterochromia

Different colors of the iris

May indicate intraocular disease.

Examples:

- Green tinge in intraocular hemorrhage.
- Brown in Siderosis resulting from a retained intraocular foreign body.



Brushfield's Spots

A series of pigmented nodules arranged around the iris.

In Down's syndrome (not specific).

Lisch Nodules



Iris hamartomas (Brown neurofibromatous nodules)
In Neurofibromatosis type I.
Seen on slit lamp / not by naked eye.

Coloboma (plural Colobomata)

Congenital anomaly (defect).

- Some of the structures of the eye are absent due to incomplete fusion of the foetal intraocular fissure during gestation / early part of pregnancy.



Typical Coloboma:

- Located in the infero-nasal quadrant.
- Affects any part of the globe from the iris to the optic nerve.
- Often associated with microphthalmia.
- Idiopathic or associated with various syndromes.
- Complications vary, depending on the location and size of the colobomata: If the defect extends further back in the eye there may be some loss of vision.

keyhole-shaped

Aniridia



- Absence of [iris](#) due to extreme hypoplasia.
- Rare [congenital](#) condition.
- Usually occurs in both eyes.
- Could be an isolated finding or in association with ocular defects (*poor development of the [retina](#) resulting in [visual loss](#)*) or in association with systemic defect (*Wilms tumor, genitourinary abnormalities and mental retardation*).

Telangiectasia

Permanent dilation of pre-existing blood vessels (*capillaries, arterioles, venules*), creating small focal red lesions, usually in the skin or mucous membranes.

Skin Telangiectasia



In **Ataxia Telangiectasia** (*an autosomal recessive disorder*), telangiectasia, cerebellar ataxia and mixed humoral and cellular (IgA, IgG, IgE) deficiency and lymphopenia are important features.



Sub-Conjunctival Hemorrhage

Benign as in vaginal delivery ; bleeding tendency.



Red Conjunctivae

Some patients with migraine particularly migrainous neuralgia develop a **red eye** on the affected side at the time of the attack.

Lens

Normal lenses are not visible by naked eyes, instead a red reflex (*orange, red or yellow*) is seen in both eyes on ophthalmoscope.

The red reflex is an extremely important screening tool.



Leukocoria refers to a white reflex instead of red one as in cataract. Sometimes could be seen by naked eye.

Urgent evaluation is a must. Retinoblastoma should be considered.



Cataract: could be:

- Congenital or acquired
- Unilateral or bilateral

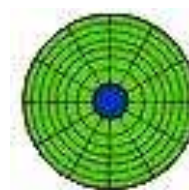
Pupils



Normally are round, regular, and nearly equal in size.

Pupil Size

The amount of light to enter the eye depends on the size of the pupil, which is controlled by the surrounding iris where the sympathetic (acetylcholine) & parasympathetic (noradrenaline) nerves act on different parts of the iris.



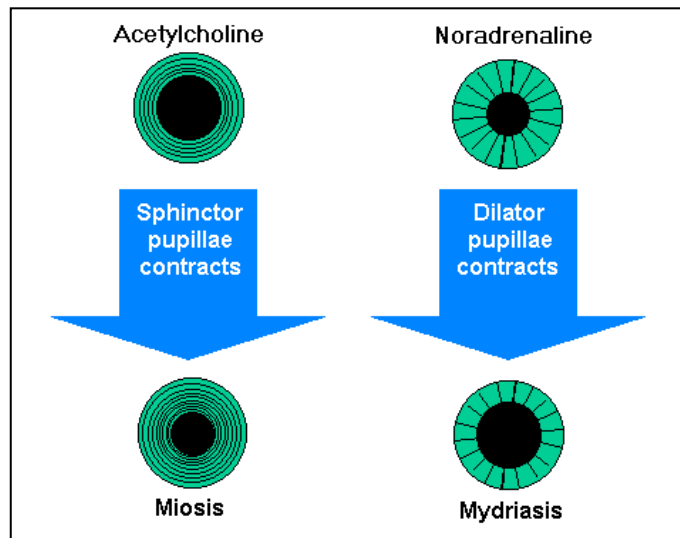
Circular Fibers
(Constrict)



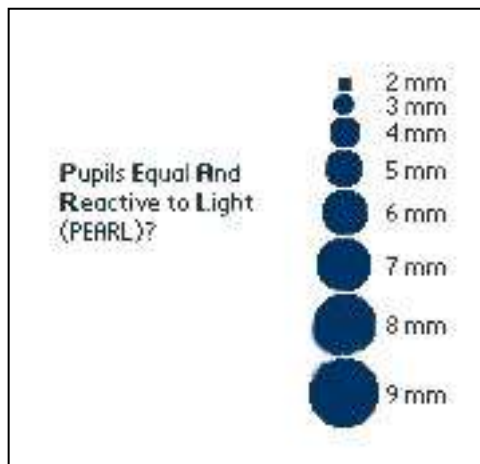
Radiating Fibers
(Dilate)

The iris contains two layers of smooth muscle:

- Sphincter pupillae (muscle fibers arranged concentrically around the pupil)
- Dilator pupillae (muscle fibers arranged radially around the pupil).



Normal Pupil Size



Not less than 3.0 mm to not more than 6.5 mm under any of following 3 different conditions in which the pupil size is measured:

- Room light.
- Near-total darkness.
- Direct light.

Pupil sizes above or below these values in these conditions may be considered abnormal.

Miotic in:

- Infancy
- When exposed to bright light
- Sleep
- Convergence
- General anesthesia and morphine addicts
- Horner's syndrome
- Oculomotor nerve palsy



Mydriatic in:

- Excitement
- Anxiety
- Dark.

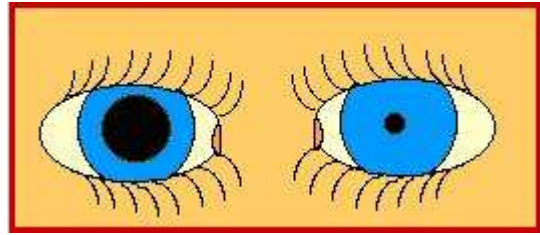


Anisocoria

A condition characterized by an unequal size of the pupils.

Physiological Anisocoria:

- A difference of 1mm or less is commonly seen in normals (*in about 20% of people*)Inherited Physiologic Anisocoria
- Eye-drops in one eye.
- Could be transient condition.

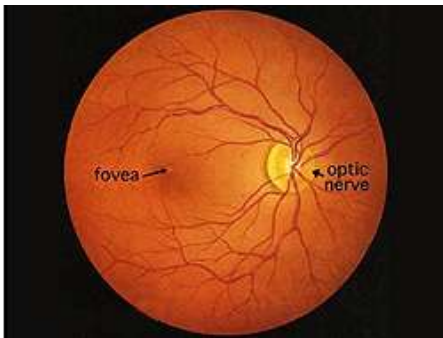


Pathological Anisocoria:

- An indication of an underlying syndrome:
 - Head injury / Intracranial bleeding.
 - Abscess or infections.
 - Expanding brain lesion / Brain tumor, aneurysm.
 - 1st sign of brain stem herniation in brain death.
 - Excess pressure in one eye / glaucoma.
 - Horner's syndrome / *drooping eyelid on the affected side with constriction of the pupil and decreased sweating on the affected side.*



Visual Fixation



An eye positioning that causes the image of an object to focus at the **fovea** (*the part of the retina which contains the highest density of cones and therefore has the best visual acuity*).

A normal child fixates on an object and follows it moving.



Questions:

- Does she/he fixate.
- Is there eye-to-eye contact / sustained eye contact.

Amblyopia (lazy eye)

A disorder of the eye that is characterized by poor or blurry vision in an eye that is otherwise physically healthy and normal. Not correctable with eye-glasses. No known organic cause.

Testing for Amblyopia:

- Put the child in parent's lap.
- Present interesting object at arms length (examiner's face or penlight, toys or bright colors).
- Move object through child's fields of gaze.
- Repeat procedure with one eye covered.
- Avoid frightening child when covering eye.

Interpretation:

- *The amblyopia is present if the child consistently cries when the good eye is covered.*
- *The amblyopia is not present if the child doesn't protest when the amblyopic eye is covered*

NB. If the child with a physically healthy eye doesn't fixate and doesn't follow objects, think of **cortical blindness**.

Ocular Movements

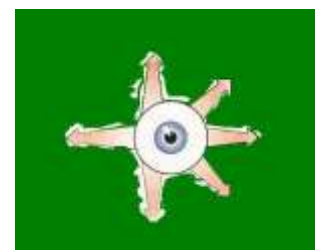
Normal eye Movements:

- **Conjugate eye movements:**

Are those that preserve the angular relationship between the right and left eyes. For example, when you move both eyes left and then right, a conjugate movement is made. Up and down movements and combinations of vertical and lateral movements also fall into the conjugate category.

- **External Ocular Movements:**

6 cardinal directions (up ; down ; medial ; lateral ; infero-lateral ; superior-lateral) / using a cross or "H" pattern.



- **Vergence eye movements:**

Are ones where the angle between the eyes change. As you move your finger closer to your face both eyes will converge and as you move your finger away from your face your eyes will diverge.

Observe for:

- Conjugate or Dysconjugate.
- Abnormal eye movements.

Squint / Strabismus

The eyes are normally parallel in all positions of gaze. If they are not, then a squint is present.

Squint could be paralytic or non paralytic.

Paralytic Squint

Associated with paresis of one of the extraocular muscles. Usually acquired.



It causes diplopia (double vision).

If longstanding or congenital, may cause abnormal head posture to minimize diplopia.

To examine for diplopia: we test the movements in all positions of gaze, the eyes should move symmetrically with no diplopia. If diplopia is present then the most peripheral double image is the one from the paretic eye.

Non Paralytic Squint

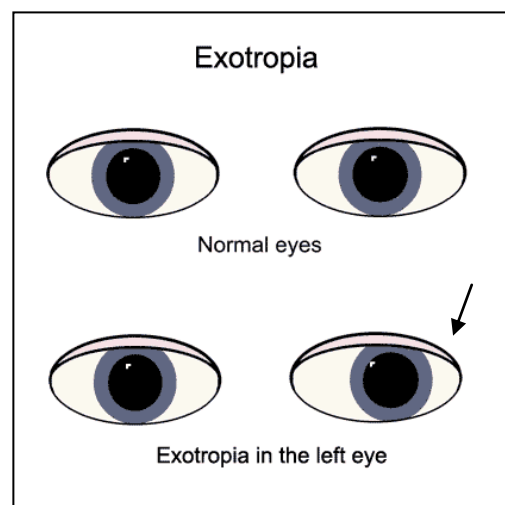
Caused by imbalance in ocular muscle tone, usually manifest in childhood. May be hereditary.

It is not associated with diplopia (because this symptom is suppressed centrally in young children), but with amblyopia.



Classified according to direction into:

1. Convergent strabismus (esotropia)
2. Divergent strabismus (exotropia)



To Examine for Squint

- **Observe the corneal light reflex:**

A light held at about 1m should produce a reflection in the center of the pupil (or slightly nasal to the center).

If the reflection in one eye is at a different location than the other, then there is probably a squint.

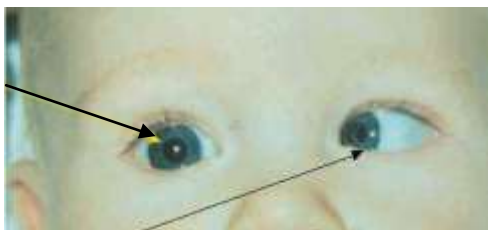


- **Perform the cover test:**

The patient looks into the distance (use a toy for a child), then we cover one eye and observe the movement of the other eye, if it moves to take up fixation then that eye was squinting. We repeat for the other eye.



Hirschberg Reflex



Note that the light reflexes do not fall in the same place in each eye. This is called the Hirschberg test. As the child is looking at you with his right eye, the examiner's light falls close to the centre of the right pupil.

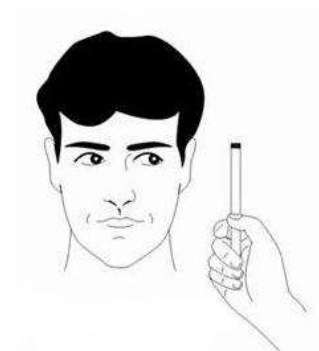
However, since the left eye is crossed, the light reflex falls on the left iris temporal (lateral) to the pupil. This is a screening test for the recognition of strabismus.

Nystagmus

A rhythmic oscillation of the eyes / unilateral ; bilateral

Types: *(based on the direction of the fast component)*

- Spontaneous / physiological or cerebellar
- Horizontal / either leftward or rightward / labyrinthine, cerebellar, brainstem or medication toxicity.
- Vertical / cerebellar or brainstem pathology. Medication toxicity
- Pendular / no fast component / blindness, albinism,
- Chaotic / dancing eyes (opsomyoclonic Syndrome)



Check for nystagmus:

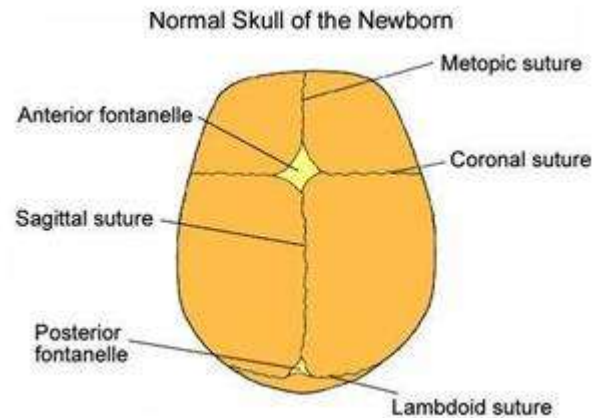
- Stop short of the extremes of lateral gaze, to keep the stimulus within the visual field of the adducting eye.
- Is performed during the examination of eye movements.

The Head

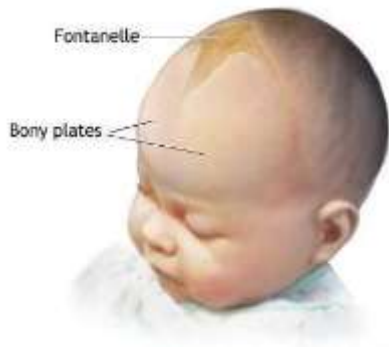
Fontanels

- An infant has 2 fontanels at birth:
 1. Anterior
 2. Posterior

Serration سن منشار Sutures are recognizable as radiolucent linear bands running to and from the wide-open fontanels



Anterior Fontanel



- Diamond-shaped
- Situated in the midline at the junction of the coronal and sagittal sutures.
- Varies greatly in size, but the usual measurement approximates 2 by 2 cm.
- The average upper limit of closure is 18 months.
- May normally close as early as 6-9 months.
- A very small or absent

anterior fontanel at birth may indicate premature fusion of the sutures or Microcephaly.

- A very large fontanel could signify a variety of problems (growing hydrocephaly, ..).



Posterior Fontanel

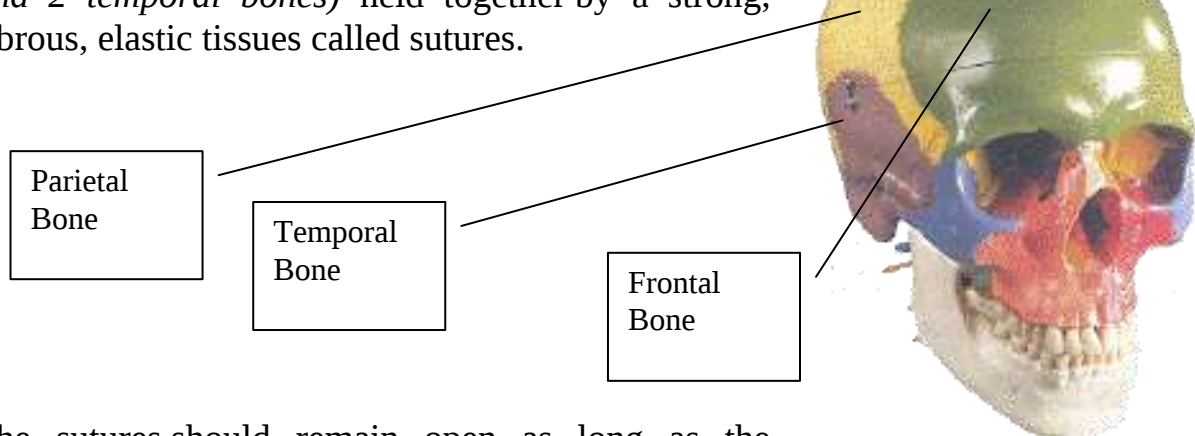
- Between the intersection of the occipital and parietal bones.
- Normally may be closed at birth or, at the most, admit the tip of a finger.
- Closes and became non-palpable after the first 6-8 wk. of life.
- Beyond this age its persistence is abnormal and is suggestive of underlying hydrocephalus or the possibility of congenital hypothyroidism.

The fontanel is normally slightly depressed and pulsatile. Best evaluated when an infant is held upright while asleep or feeding.

A bulging fontanel is a reliable indicator of ICP or hydrocephalus. But vigorous crying can cause a protuberant fontanel in a normal infant.

Cranial Sutures

The infant skull consists of 6 plates of bone (*the frontal bone, the occipital bone, 2 parietal bones, and 2 temporal bones*) held together by a strong, fibrous, elastic tissues called sutures.



The sutures should remain open as long as the brain continues to grow, enabling the skull to expand & properly accommodate the brain's growth.

When a suture closes prematurely, a predictable abnormality of head shape occurs. The spaces between the bones where the sutures lie are called fontanel (the anterior fontanel and the posterior fontanel).

The cranial bones remain separate for approximately 12-18 months. They then grow together (fuse) and remain fused throughout adulthood.

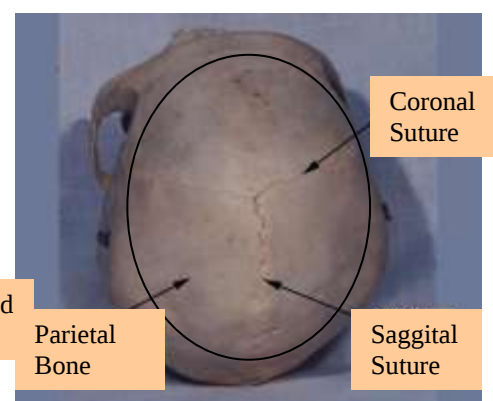
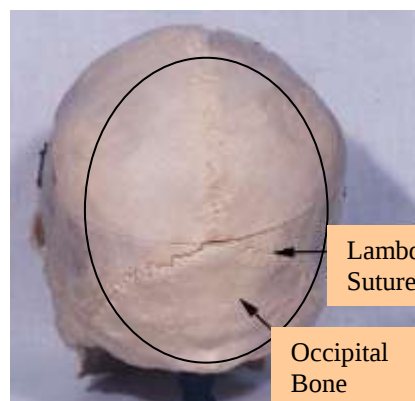
The fibers (sutures) and spaces between the cranial bones (fontanel) are necessary for the infant's brain growth and development. During childbirth, the flexibility of the fibers allows the bones to overlap their edges so the head can pass through the birth canal without compressing and damaging the infant's brain.

During infancy and childhood, the flexibility of the fibers allows the rapid growth of the brain without constriction.

Feeling the cranial sutures and fontanel is one way that physicians determine the child's growth and development.

Cranial sutures:

1. Coronal
2. Metopic
3. Sagittal
4. Lambdoid



The size of the sutures varies widely at birth and they can be from 1.5 mm to 10 mm. The wider sutures narrow rapidly and most are below 3mm within a few months.

Timing of Closure of Sutures and Fontanel's

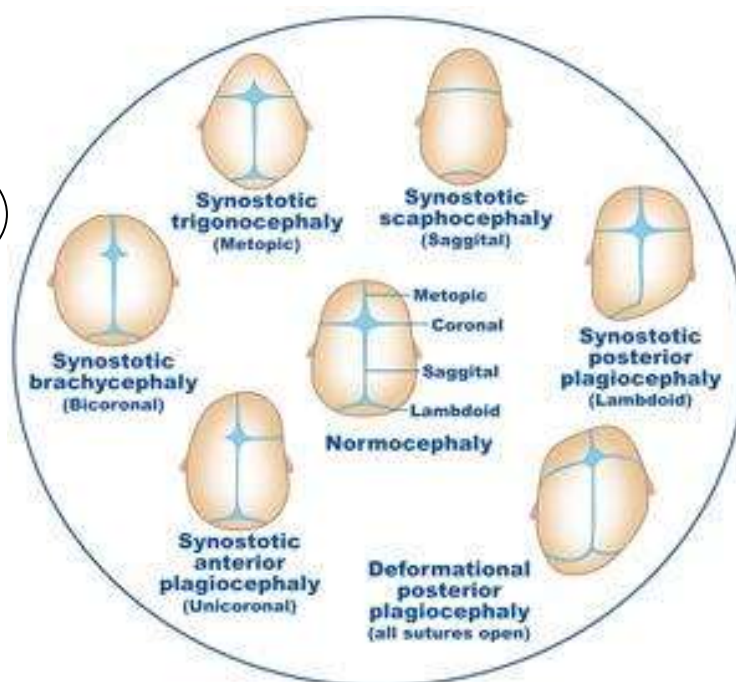
Type of Suture/ Fontanel	Time to Closure
Metopic suture	9 months-2 years (may persist into adulthood)
Coronal, sagittal, lambdoid sutures	40 years
Anterior fontanel	9 - 18 months
Posterior fontanel	3 – 6 months

Craniosynostosis

Premature fusion of one/or more suture/s.

Cranio /skull ;
Synostosis / fusion of
open areas (sutures)

If occurs, skull growth can't occur perpendicular to that suture but continues to occur in the line parallel to the fused suture (eg. *sagittal fusion results in scaphocephaly*).



Types of Craniosynostosis

Metopic Synostosis

The metopic suture is located between the frontal bones and the nose. Early closure of this suture results in trigonocephaly. Infants will often have a prominent ridge down the middle of the forehead, a pointed forehead, and eyes that seem too close together creating a cosmetic problem only.



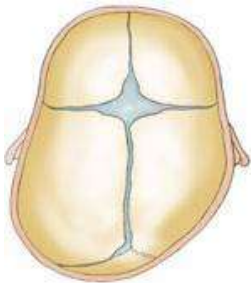
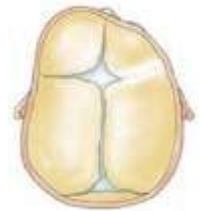


Sagittal Synostosis

Early closure of the sagittal suture results in scaphocephaly. Infants will often have a protruding forehead, an elongated head, and a bony protuberance at the back of the skull.

Coronal Synostosis

Coronal synostosis may occur on either side or may be bilateral. Infant will often have an elevation of the eye socket تجويف, flattening of the ridge of the eye and displacement of the nose on the affected side.



Lambdoid Synostosis

Lambdoid synostosis is rare. It may occur on either side or may be bilateral. Infants will often have flattening of the back of the skull on the affected side.

Head Shapes

The neonate skull may appear elongated and sometimes shows overlapping of cranial bones from the moulding during birth. These appearances change to a more contour within a week or so.

Plagiocephaly

- Asymmetry and twisting configuration of the head.
- From the above appears as parallelogram.
[متوازي الاضلاع]
- Most often a 'postural' deformity which corrects spontaneously once the child is mobile.
- Unilateral coronal synostosis.



Scaphocephaly = Dolichocephaly

- The head is long in the antero-posterior diameter and narrow when viewed from the front due to Sagittal synostosis.
- Usually associated with prematurity.
- Sometimes Hurler's syndrome.



Turricephaly = Acrocephaly = Oxycephaly



- A cone shaped head / Tower-head
- The head is tall due to compensatory upward growth due to premature bilateral coronal synostosis.
- Raised intracranial pressure is especially likely to occur with this deformity.
- Think of Alpert's syndrome and Carpenter's syndrome.

Brachycephaly

- A short head, one that is short in diameter from front to back.



- The word brachycephaly comes from the Greek brachys = short ; kephale = head.
- The back of the head is flattened due to restricted growth of the forehead in a forward direction mainly when there is early bilateral coronal synostosis.



Trigonocephaly



- The head is triangular in shape with narrow pointed forehead due to premature in utero fusion of the metopic suture.
- The main problem is cosmetic.

Anencephaly

- A fetal abnormality in which there is no development of the forebrain. The cranial vault is absent, and the anterior pituitary gland may be absent or hypoplastic.
- It is due to a failure of closure at the cephalic end of the neural tube.
- Survival is limited. If live-born, survive only a few hours.
- The risk of recurrence of this condition is about 1:30. This rises to about 1:10 if two previous children have been affected.



Head Palpation

- The newborn's skull characteristically shows overriding of the cranial sutures for the first several days (*pressures exerted on the skull during its descent*). After few days should arouse suspicion.
- Sutures are easily felt in the newborn infant.
- **Craniotabes** (*softening of the parietal bone*): gentle pressure produces a sensation similar to indenting a ping-pong ball.

Head Auscultation

- Cranial Bruits are best heard by the diaphragm of the stethoscope.
- Most prominent over the anterior fontanel, temporal region, or the orbits:
 - In febrile illness.
 - Arterio-venous malformations
 - Heart murmurs → transmitted to the cranium.
 - Severe anemia
 - Increased intracranial pressure → significant intracranial bruits.

Head Circumference (HC)

Occipito-frontal circumference

Measuring HC is an accurate reflection of brain size. The brain grows to 80% of its adult volume during the first 2 years of life, so many neurological diseases that occur early in life will impact the growth of the brain.



Technique of measurement:

- Non distensible plastic measuring tape is placed over the mid-forehead and is extended circumferentially to include the most prominent portion of the occiput, so that, the greatest volume of the cranium is measured.

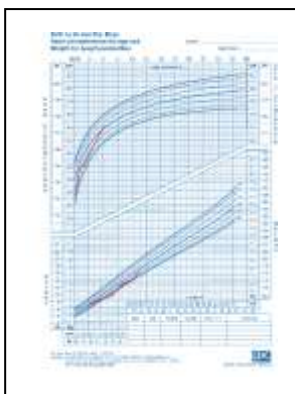
Factors producing error in measurements:

- Scalp edema
- Overriding of the sutures
- Intravenous fluid infiltration
- Cephalohematoma



Considerations:

- In newborn the HC > Chest circumference by about 2 cm.
- 6 months - 2 years both measurements are about equal.
- After 2 years, the chest C. > HC.



It is essential to plot head circumference on a standardized head growth chart.

A series of measurements over time that show an increased rate of head growth often can provide more valuable information than a single measurement that is larger than expected. However, in some cases a single measurement is sufficient to confirm a significant large head.

Average Rate of Head Growth

- **Premature:**
 - 0.5 cm in the 1st 2 wks.
 - 0.75 cm during the 3rd wk.
 - 1 cm in the 4th wk. (*40th wk. of gestation*).
- **Full term:**
 - At birth 34-35 cm
 - By 6 months 44 cm
 - By 1 yr. 47 cm

What the Percentiles Mean

- A standard growth chart is composed of seven curves that follow the same pattern. Each curve represents a different percentile: 5th, 10th, 25th, 50th, 75th, 90th, and 95th.
- The child's measurements will be plotted on the charts, so the comparison to the average for age group can be done and helps to determine whether the child is growing as expected.
- **Interpretation:**
 - The 50th percentile line represents the average for a particular age group.
 - The 90th percentile means that the measurement is greater than 90% of children of that age in the country. Only 10% of infants of that age have measurements exceeding those of this infant.
 - If a 4-year-old's weight at a checkup is at the 20th percentile, that means about 80% of children that age weigh more than that child, and 20% of kids in the United States weigh less than that child.
- Just because a reading is high or low on the chart doesn't necessarily mean that there's a problem. A baby whose head circumference is in the 90th percentile might also fall in the 90th percentile for weight and length, which would mean that the infant is just a normal kid who's large overall. By the same token, the child whose weight is at the 20th percentile may have parents who are a bit smaller than average for height and weight. For that child, being in the 20th percentile is an entirely normal reading, as long as he or she is growing fairly steadily along that percentile curve over time.
- Sometimes, however, a child's measurement increases or falls to a higher or lower percentile curve or is at one extreme of the growth chart. For example, a child who falls below the 5th percentile on the weight for height would be considered underweight and a child who is at or above the 85th percentile would be considered at risk for becoming overweight; those at or above the 95th percentile are considered to be overweight.

Don't focus too much on any one reading. Instead, the numbers should be viewed as a trend. Any measurement, taken out of context from the others, might give you the wrong impression.

When growth chart readings are examined over time, they reveal a pattern of growth. That pattern reflects how the child is growing in relation to other children of his or her age and also shows the progression from previous measurements. This information is a much more useful indicator of whether a child is growing normally than any single measurement.

Macrocephaly

Occipito-frontal circumference > 2 standard deviations above the mean for age, sex, height and weight.

Increased intracranial pressure usually caused by swelling of the brain or accumulation of cerebrospinal fluid within ventricles) often accompanies increased head circumference. Symptoms associated with this include vomiting, eyes deviating downward, and irritability.



Common Causes of macrocephaly:

- Hydrocephalus (congenital, post-traumatic, or obstructive).
- Benign familial macrocephaly (family tendency toward large head size).
- Metabolic disorders (Canavan disease, Alexander, Hurler syndrome, Morquio syndrome).
- Intracranial bleeding.

Medical history questions, documenting increased head circumference in details, may include:

- Time pattern
 - When first noticed that the baby's head seemed large?
 - Does the baby's head size seem to be increasing faster in proportion to the growth of the body?
- Location
 - Does the head seem larger all over?
 - Is the head growing more in a front-to-back pattern or in a side-to-side pattern?
- Other
 - What other symptoms are also present (especially changes in brain or nervous system functions)?



Hydrocephaly, mainly the progressive one is managed mostly by VP-Shunting

Microcephaly / small head

Occipito-frontal circumference > 2 standard deviations below the mean for age, sex, height and weight.

1. Primary microcephaly

- Autosomal recessive primary microcephaly (MCPH).
- Autosomal dominant primary microcephaly.
- X-linked primary microcephaly.
- Syndromic primary microcephaly
 - Fanconi anemia
 - Cockayne syndrome
 - De Lange syndrome
 - Meckel syndrome
 - Fetal akinesia syndrome with lissencephaly type III
 - Chromosomal diseases.
 - Wolf-Hirschhorn syndrome (4p- deletion).



2. Secondary microcephaly (environmental microcephaly)

- Prenatal infections.
- Drugs.
- Chemical agents.
- Intrauterine growth retardation (IUGR).

3. Associations

- Microcephaly and acidosis.
- Amish type microcephaly.

Mucous Membranes & Skin Examination

Neurocutaneous Syndromes

Telangiectasia



Telangiectasia are dilated superficial tiny, red "spider" veins which appear in the corners of the eyes or on the surface of the ears and cheeks.

Are characteristic of **ataxia-telangiectasia**, but are not always present and generally do not appear in the first years of life.

Ataxia Telangiectasia (A-T) is a rare, progressive, neurodegenerative childhood disease that affects the brain and other body systems.

Telangiectasia, ataxia and immunodeficiency are the diagnostic triad.

About 20% of those with A-T develop cancer, most frequently acute lymphocytic leukemia or lymphoma.

Children with A-T usually have normal or above normal intelligence.



Hypopigmented Macules

Ash-Leaf Depigmentation [ورق نبات رمادي]

Shagreen Patch [جلد غیر مدبوغ / شامواه]



Seen in Tuberous sclerosis



Café-au-Lait Spots (*Neurofibromatosis*)

More than 5 café au lait patches are significant.



Hemangioma / Port-wine Stain

Area of one or more divisions of the Trigeminal nerve, usually the ophthalmic division

In Sturge Weber Disease



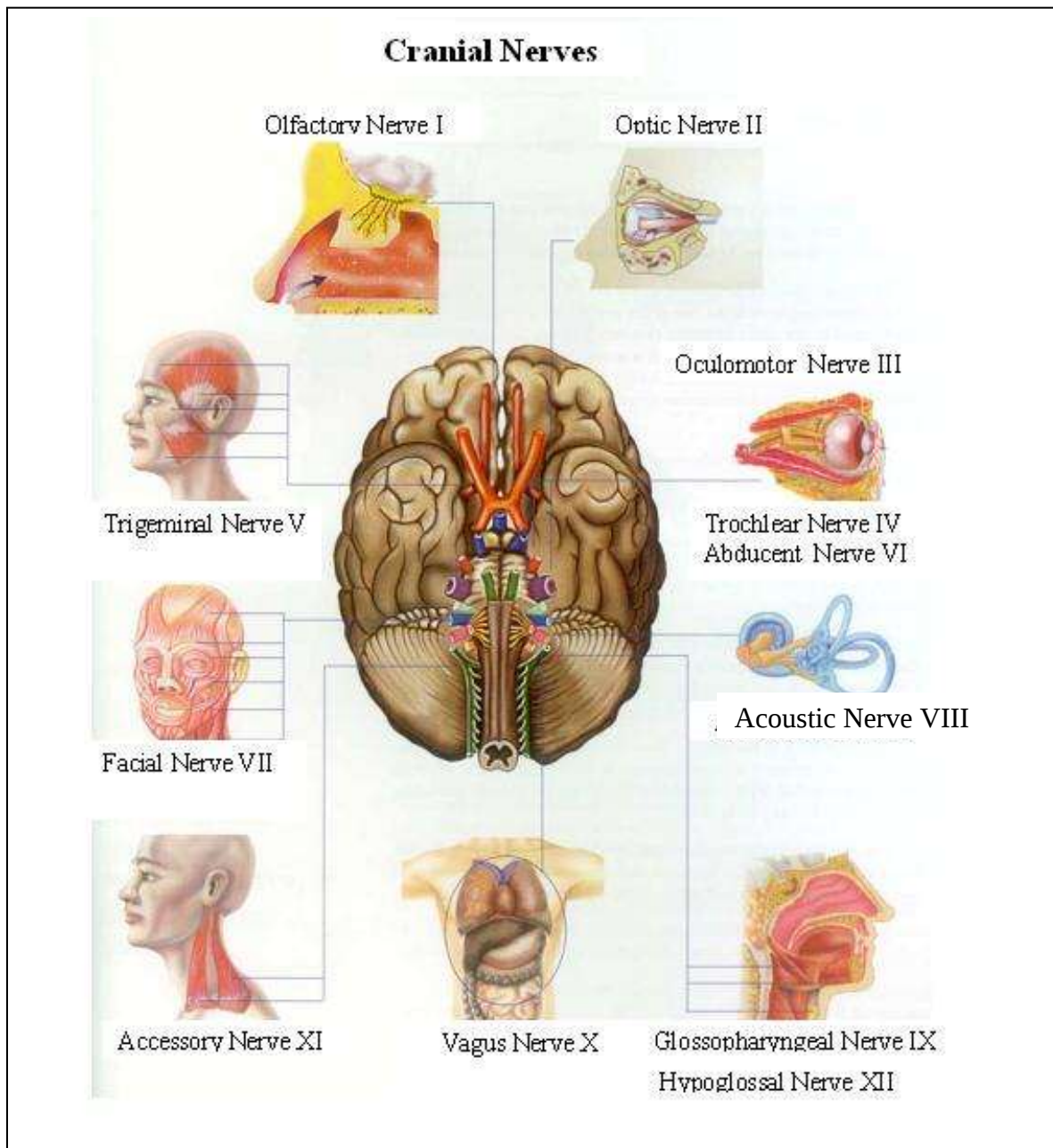
Cranial Nerves

12 pairs of cranial nerves (CN):

- 1st Olfactory
- 2nd Optic
- 3rd Oculomotor
- 4th Trochlear

- 5th Trigeminal
- 6th Abducent
- 7th Facial
- 8th Acoustic

- 9th Glossopharyngeal
- 10th Vagus
- 11th Accessory
- 12th Hypoglossal



General Considerations

CNs are attached to the brain and transmitted through foramina in the base of the cranium and lead directly to various parts of the head and neck.

They emerge from, or enter the cranium, as opposed to the spinal nerves which emerge from the vertebral column.

The area of attachment to the surface of the brain is termed “**superficial or apparent origin**”.

The **motor (efferent)** cranial nerves arise within the brain from groups of neurons which constitute their “**nuclei of origin**”.

The **sensory or (afferent)** cranial nerves arise from groups of neurons outside the brain that may be grouped to form ganglia on the trunks of the nerves or may be situated in peripheral sensory organs such as the nose and eye. The central processes of these cells run into the brain, and there, end by arborizing around neurons, which are grouped to form “**nuclei of termination**”.

The **nuclei of origin** of the motor nerves and the **nuclei of termination** of the sensory nerves are brought into relationship with the cerebral cortex after crossing the middle line to the opposite side.

- Remember that a deficit found may reflect pathology in central pathways, rather than in the nerve itself or a problem in the muscles (or receptors) that the nerve innervates.
- Some are limited to motor and some to sensory functions. Some are mixed and others are specialized (smell, vision, hearing).

Abnormalities	Affected Cranial Nerves
Ptosis	III
Facial Droop (متدلي) or Asymmetry	VII
Hoarse Voice	X
Articulation of Words	V, VII, X, XII
Abnormal Eye Position	III, IV, VI
Abnormal or Asymmetrical Pupils	II, III

Examination of Cranial Nerves in Newborn

Examination of the baby's cranial nerve function is often accomplished by observing spontaneous activity:



- During crying, facial movement (Cranial Nerve VII) is observed for fullness or asymmetry.
- The quality and strength of the cry is a way of looking at Cranial Nerves IX and X function.
- Sucking and swallowing assesses Cranial Nerves V, VII, IX, X, and XII because all of

these cranial nerves are involved in this complex act.

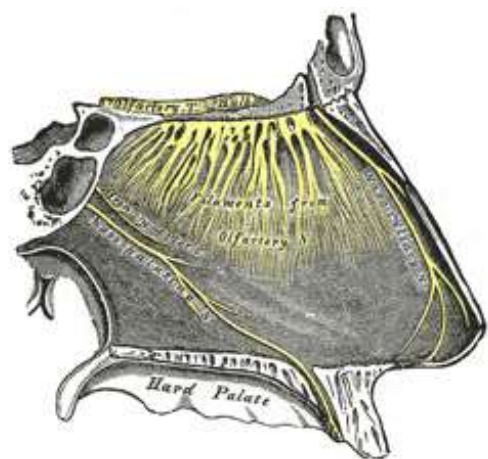
- Eye movements (Cranial Nerves II, IV and VI) can be assessed by using the vestibulo-ocular reflex (doll's eyes maneuver). When the head is turned, there is conjugate eye movement in the opposite direction.
- Testing a baby's behavior response to light (Cranial Nerve II).
- Testing a baby's behavior response to sound (Cranial Nerve VIII).
- Pupillary light reflex, corneal reflex, gag reflex and fundoscopic exam are done in the same manner as the adult exam.

Olfactory nerves (I)

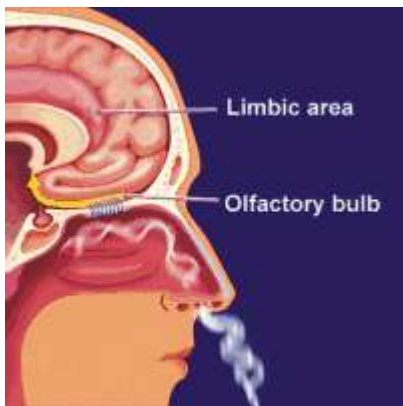
Olfactory Special Sensory: Smell

Generalities

- The word "olfactory" comes from the Latin "olfacere", to smell.
- Register smells by carrying the impulses, for the sense of smell, from the nose to the brain. The olfactory nerves are the only nerves in the human body known to regenerate continually through all of childhood and adulthood (self-renewal).
- The olfactory system consists of the olfactory epithelium (olfactory region of the nasal cavity: the superior nasal concha, and the corresponding part of the nasal septum), bulbs and tracts along with olfactory areas (olfactory center) of the brain collectively known as the rhinencephalon. Have only a special sensory component (special afferent).



Functions



- Sense of smell حاسة الشم
- No efferents (no motor).

Pathology

- The sense of smell is not highly developed in mankind and is easily disturbed by conditions affecting the nasal mucosa (e.g. common cold).

Clinical Signs

- Loss of ability to smell

How to Examine

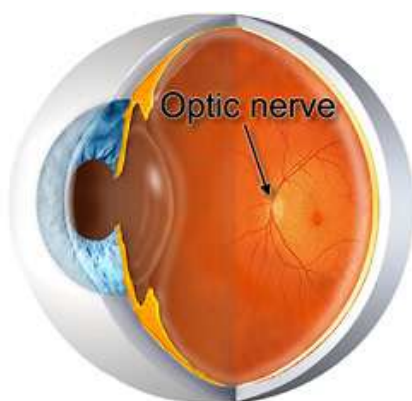
- Not routinely Tested
- Use familiar and non-irritating odors (known aromatic substance).
- Assure that the patient can move air through each nostril.
- Close both eyes.
- Ask to sniff in one nostril / while occluding the other.
- Ask to detect the presence or absence of the odors (held near the nose, or withdrawn).

Optic nerves (II)

Optic Special

Generalities

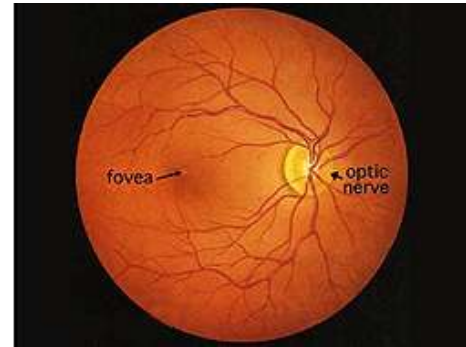
- The optic nerve acts like a cable connecting the eye with the brain. It is more like brain tissue than it is nerve tissue.



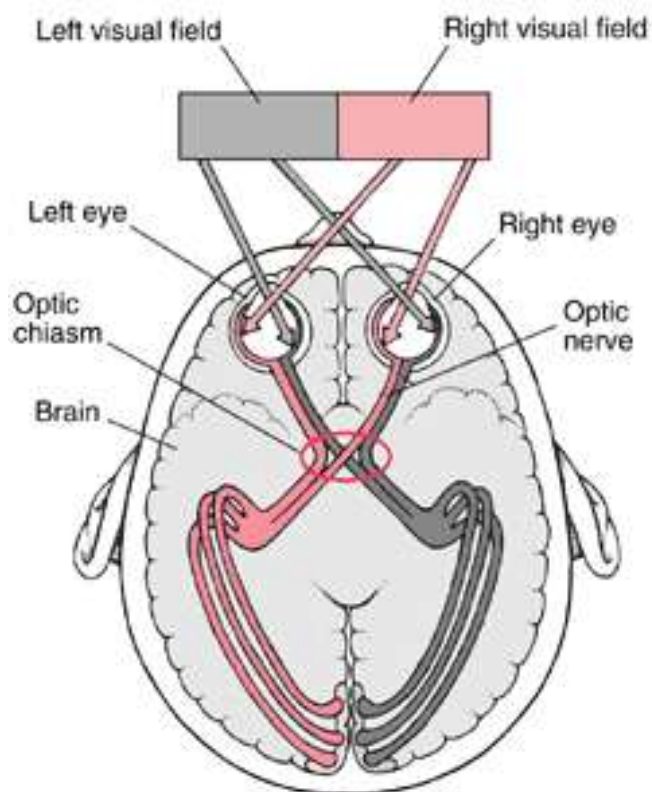
- The optic nerves, Rt. and Lt., are composed primarily of axons from the retinal ganglion cells. Each one transmits electrical impulses from the retina to the brain. It begins at the optic nerve head (disc), near the macula, and passes through the orbit and optic canal to the chiasm then through the optic tracts passes to the cortical visual center situated in the occipital lobe.
- A few fibers pass as afferent fibers (pupillary reflexes).

- The optic nerve orbital portion is 20-30 mm, while the inter-cranial portion is 10 mm.

- **Central vision:** Straight-ahead vision. Is the work of the macula. Central vision permits a person to read, drive, and perform other activities that require fine, sharp, straight-ahead vision (reading, watching television or movies, driving, and any activity where visual detail is of primary importance), as opposed to peripheral vision.



- The retina is made up of two types of cells, the cones and the rods. Millions of cones are packed into the macula. The cones are nerve cells sensitive to light, fine detail, and color. The **macula** is an oval yellow spot near the center of the retina of the human eye. It has a diameter of about 1.5 mm and contains a rich collection of cones. Near its center is the **fovea**, a small pit that contains the largest concentration of cone cells in the eye and is responsible for central vision. It is specialized for high acuity vision.
- The retina's sensory receptor cells are absent from the optic nerve. Because of this, everyone has a normal blind spot. This is not normally noticeable because the vision of both eyes overlaps.



- Within the chiasma, the optic nerves undergo a partial **decussation**. Its principal part consists of two sets of fibers: **crossed** and **uncrossed**.
- The crossed fibers (more numerous / fibers emanating from the nasal half of each retina), occupy the central part of the chiasma, and pass from the optic nerve of one side to the optic tract of the other, decussating in the chiasma with similar fibers of the opposite optic nerve. The uncrossed fibers (originating in the temporal retina) occupy the lateral part of the chiasma, and pass from the nerve of one side into the tract of the same side.
- The chiasma is located just below and in front of the pituitary gland (which is why a tumor on the pituitary gland, pressing on the optic chiasm, can affect vision).
- The optic nerves are surrounded by a three-layered meningeal sheath similar to those of the CNS which consists of a dura (optic nerve sheath), arachnoid, and pia. The subarachnoid space is in direct communication with the subarachnoid space of the CNS. These sheaths are separated from each other by cavities which communicate with the subdural and subarachnoid cavities respectively.

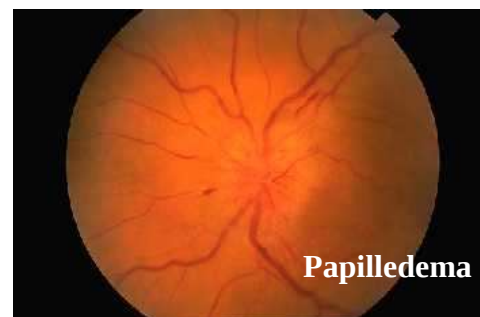
Clinical Signs

Signs of optic nerve degeneration:

- Visual field defects
- Blood vessel changes around the optic disc
- Optic disc pallor
- Pupil reflex abnormalities
- Color vision abnormalities

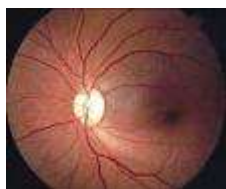
Papilledema

A swelling or edema of the optic disc. Histopathologically, the axons in the optic disc are markedly edematous and swollen with surrounding tissue edema and also increased vascular congestion



Most commonly it is bilateral, secondary to an increase in intracranial pressure. Vision is not affected initially. Secondary optic atrophy and permanent vision loss can occur if the primary cause of the papilledema is left untreated.

Optic Atrophy



Optic atrophy means the loss of some or most of the nerve fibers in the optic nerve resulting in little visual change to severe visual loss.

The optic disc will appear pale or white, indicating loss of nerve fibers.

- It can be congenital (usually hereditary) or acquired (vascular, degenerative, pressure, metabolic, trauma, glaucoma, or toxicity of the nerve fibers of the optic nerve and optic tract).
- Loss of vision is the end result.
- A pale optic disc and loss of pupillary reaction are usually proportional to the visual loss.
- Degeneration and atrophy of optic nerve fibers are irreversible.

Optic Neuritis

- Is an inflammation of the optic nerve.
- It may affect the part of the nerve and disk within the eyeball (papillitis) or the portion behind the eyeball (retrobulbar optic neuritis, causing pain with eye movement).
- There will be no visible changes in the optic nerve head (disk) unless some optic atrophy has occurred.
- Can be caused by demyelinating diseases (e.g., multiple sclerosis, post-infectious encephalomyelitis), systemic infections, nutritional and metabolic diseases, Leber's Hereditary Optic Neuropathy.
- The condition is unilateral rather than bilateral.



Efferent Pupillary Defect

- Seen in Oculomotor nerve palsy and parasympathetic blocking drops.
- Paralysis of Oculomotor nerve (CN III) gives mydriasis (dilated pupil).
- No reaction to light or accommodation.
- The other pupil reacts consensually.
- Associated findings: ptosis, diplopia, squint.

Afferent Pupillary Defect (Marcus Gunn pupil)

- Seen in optic nerve disease.
- Poor direct light reaction, normal consensual reaction and accommodation on the affected side, consensual reaction on the normal side is reduced.
- The abnormal pupil is dilated.



Horner's Syndrome (HS)

- Seen in sympathetic lesion.
- Normal reaction to light and accommodation
- Does not dilate with cocaine drops or in dark
- The pupil is constricted
- Associated findings: ptosis, impaired sweating, enophthalmos
- There is abolition of the ciliospinal reflex (dilatation of the pupil when the skin of the neck is pinched. This is due to reflex excitation of the pupil dilating fibers in the cervical sympathetic chain).

To diagnose HS, put the patient in a dark room, the normal pupil dilates much better.

Drugs & Pupils Size

Bilaterally constricted pupils:

- Neostigmine, pontine hemorrhage, morphine and organophosphorus poisoning.

Bilaterally dilated pupils:

- Anticholinergic- atropine. Sympathomimetic- amphetamine

How to Examine

1. Test Pupillary Light Reflex
2. Test Accommodation Reflex (*the near reflex*)
3. Test Visual Acuity
4. Screen Visual Fields
5. Optic Fundi Examination

1. Test Pupillary Light Reflexes

Normal pupils are round, regular, and nearly equal in size. They are miotic in infancy, old age, bright light, sleep, convergence, general anesthesia and morphine addicts.

Light reflexes are 2:

1. **Direct Light Reflex** (same pupil constricts to light).
2. **Consensual Light Reflex** (other pupil constricts as well the one exposed to light).

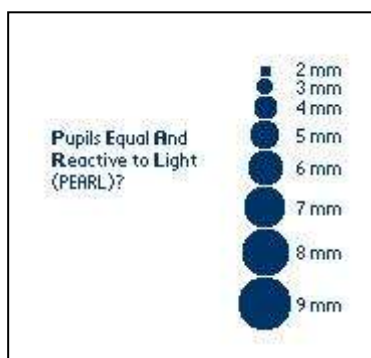
Reflex afferent pathway (sympathetic) involves the retina, optic nerve, chiasm, then optic tract with linkage between both ipsilateral and contralateral sides.

Efferent pathway (parasympathetic) pass in the Oculomotor nerve that innervate the constrictor muscles of iris.

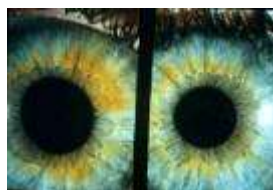
For testing pupillary light reflexes

- Dim the room lights as necessary.
- Ask the patient to fix into a distance point to avoid the accommodation response.
- Shine a bright light obliquely into each pupil.
- Look for both, the direct (same eye) and consensual (other eye) reactions.

Both pupils should **constrict equally** with **light shown in either eye.**



- Record the **speed** of constriction (*normal speed, sluggish*)
- Record pupil **size in mm** (in room light, and dim light).
- Record any **asymmetry** (anisocoria) or any irregularities of pupil shape.



Anisocoria of 1mm or less is commonly seen in normals (physiologic anisocoria).

2. Test Accommodation Reflex

When eyes converge, the pupils constrict.

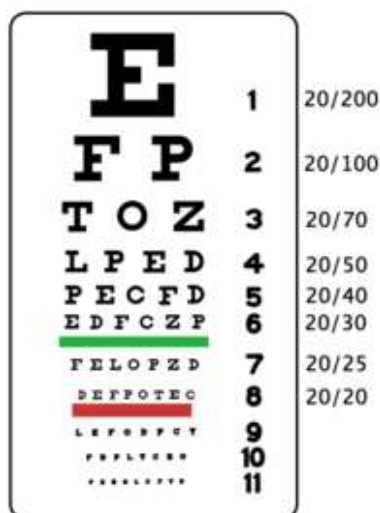
For testing accommodation Reflex:

- Have the patient shift gaze from a distant point to a point about 15 cm from the eye.
- Having the patient look at his own thumb as you move it toward his eyes provide an effective stimulus.
- See pupillary constriction (equal in both eyes) and convergence.

Interpretation:

- Normal pupils: **PERLA** = pupils equal and reactive to light and accommodation.
- Since both pupils constrict in response to light directed into one eye, afferent lesions can easily be distinguished from damage to the efferent pathway.

3. Test Visual Acuity



Visual acuity refers to the clarity or clearness of one's vision,

The word “acuity” comes from the Latin “acuitas,” which means sharpness.

Visual acuity often is referred to as “Snellen” acuity. The chart and the letters used to measure visual acuity are named for a 19th century Dutch ophthalmologist Hermann Snellen (1834–1908) who created them as a test of visual acuity.

The visual acuity test is used to measure how well a person sees. That means to determine the smallest letters a person can read on a standardized chart or card held 6 meter away.

This test is a routine part of an eye examination as undetected or untreated problems may result in permanent damage to vision.

The reason that the number “20” is used in visual acuity measurements is because, in the US, the standard length of an eye exam room (that is, the distance from the patient to the acuity chart) is about 20 feet. In England, where meters are used instead of feet, a typical eye exam room is about 6 meters long. Therefore, instead of using 20/20 for normal vision, 6/6 is used.

How the test is performed

- May be done in a health care provider's office, a school, a work place, or elsewhere.
- Stand the child behind a line 20 feet (6 meters) from the eye chart.
- Remove glasses or contacts.
- Keep both eyes open and gently cover one eye.
- Ask the child what he see.
- Perform it on each eye, one at a time.
- If necessary, it is then repeated while wearing her/his glasses or contacts.
- No special preparation is necessary for this test.

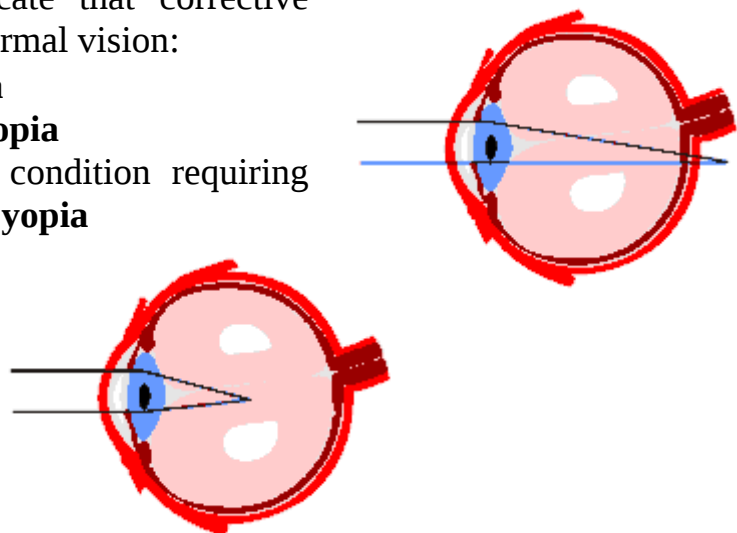
Normal Values

- Visual acuity is expressed as a fraction.
- The top number refers to the distance you stand from the chart. This is usually 20 feet (6 meters).
- The bottom number indicates the distance at which a person with normal eyesight could read the same line you correctly read.
- For example, 20/20 is considered normal. 20/40 indicates that the line you correctly read at 20 feet can be read by a person with normal vision from 40 feet away.

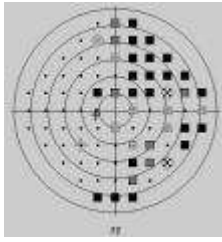
What abnormal results mean

Abnormal results may indicate that corrective lenses are needed to obtain normal vision:

- Nearsightedness = **Myopia**
- Farsightedness = **Hyperopia**
- Or may indicate an eye condition requiring further evaluation = **Presbyopia**
- **Amblyopia** = lazy eye.



4. Test Visual Field



Visual field loss, caused by optic nerve damage, is measured by using a “**Visual Field Analyzer**” or “**Perimeter**”.

Field loss usually is not even measurable until 25% to 40% of the optic nerve’s axons have been destroyed. The first fibers to die are the larger fibers, which primarily carry form and motion information, rather than the smaller fibers, which primarily detect light

5. Optic Fundi Examination (Fundoscopy)

The Fundus, or inner lining, of the eye is examined by the ophthalmoscope.

For documentation can be photographed with specially designed cameras through the dilated pupil. This painless procedure produces a sharp view of the retina, the retinal vasculature, and the optic nerve head (optic disc) from which the retinal vessels enter the eye. The optic disc measures about 1.5 mm in diameter.

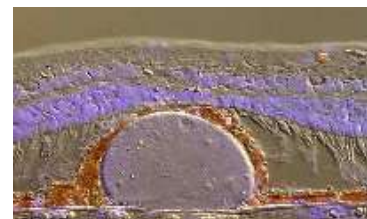
The yellowish disc is the head of the optic nerve which connects the eye to the brain. Radiating from the **optic disk** are **retinal arteries** and **veins** which supply the retina with nutrition. The vessels form an arcuate pattern delineating the macula which produces our central vision. At the center of the **macula** (in the center of above photograph) lies the tiny **fovea** which is responsible for our reading vision.



A fundus examination revealing a swollen optic nerve head with elevation and edema.



Retinal hemorrhage



Drusen
a marker for macular degeneration

Eye Movements

- Normal eye movements
- Abnormal eye movements
- Conjugate or Dysconjugate
- Abnormal eye movements

Muscles of the orbit

1. Levator Palpebrae Superioris

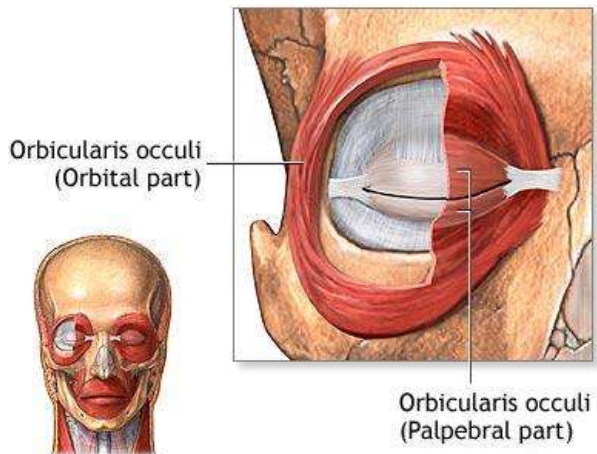
Innervation:

Oculomotor nerve

The muscle contains smooth muscular fibers innervated by sympathetic nerves from superior cervical sympathetic ganglion of the Oculomotor nerve. Its stimulation causes further elevation of the lid.

Action:

Rises the upper lid so its paralysis leads to drooping (ptosis).



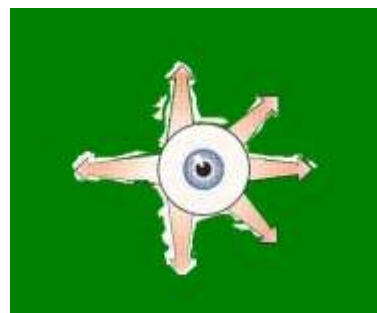
2. Recti Muscles

- The extrinsic ocular muscles (extra ocular muscles) are a series of six muscles that are attached to each eye.
- These muscles are responsible for the movement and proper alignment of the eyes.



External Ocular Movements 6 cardinal directions: "*H*" pattern

1. Up
2. Down
3. Medial
4. Lateral
5. Down-lateral
6. Up-lateral



Extrinsic Ocular Muscles		
Lateral Rectus	Rotates the eye outward toward the temple, a movement called abduction.	Abducens Nerve (CN VI)
Medial Rectus	Rotates the eye inward toward the nose (adduction).	Oculomotor Nerve (CN III)
Inferior Rectus	The primary muscle responsible for turning the eye downward, a movement called depression.	
Superior Rectus	The primary muscle responsible for turning the eye upward, a movement called elevation.	
Inferior Oblique	Rotates the eye upward and outward toward the temple, a movement called extorsion.	
Superior Oblique	Rotates the eye both downward and inward toward the nose, a movement called intorsion.	Trochlear Nerve (CN IV)

Ocular motility also depends on Supra-nuclear Connections

- Sometimes impaired movements of both eyes in one direction occurs (supranuclear gaze palsies, or conjugate gaze palsies). So loss of conjugate eye movements indicates a lesion above the level of the brainstem.
- Medial Longitudinal Fasciculus (MLF): which interconnects the nuclei, helps to keep optical axes parallel or conjugate when the eyes are turned to one side or when moving the head. Internuclear ophthalmoplegia occurs when there is a lesion of MLF. In this case the ipsilateral eye is unable to adduct and there is nystagmus in the contralateral abducting eye. This is seen most commonly in multiple sclerosis, in vascular disorders and neoplasms of the brainstem.

Oculomotor nerves (III)

Motor / Ocular movements

Generalities

Supplies somatic motor fibers to the majority of extra-ocular muscles (except the Obliquus superior and Rectus lateralis), therefore will elevate, depress and adduct the eyeball.

In addition it innervates the smooth muscles within the eye (parasympathetic neurons).

It is also responsible for:

- Pupillary light reflex (illuminating the retina causes the constriction of the pupil / the direct light reflex). Constriction of the non-illuminated eye should occur due to consensual light reflex.
- Accommodation reflex (fixation upon a nearby object by convergence of the optic axes) involves concomitant contraction of the ciliary muscle to increase the convexity of the lens thus focusing the image with pupillary constriction.

Functions



- Most extra-ocular movements (*Somatic Motor*).
- Opening of the eyes.
- Pupillary constriction.

Nerve Disorders

Isolated nerve palsy

- **Causes:** diabetic mononeuropathy, carotid posterior communicating aneurysm, tumors (pituitary and others), trauma and vascular lesions.
- **Features:** pupil dilatation, lateral deviation due to unopposed action of lateral rectus muscle, drooping and diplopia.

3rd n. palsy + contralateral hemiplegia = Weber syndrome,

- **Causes:** midbrain stroke.
- **Features:** The lesion may be incomplete depending on location and type of lesion: diabetes or vascular disease tends not to involve the pupil, but compressive lesions do (difference between medical and surgical 3rd n. palsy is the pupil dilatation).

Clinical Signs

Ptosis, mydriasis, ophthalmoplegia of adduction, depression, elevation and extorsion and diplopia.

How to Examine

- Observe for Ptosis
- Test Extra-Ocular Movements:
 - Stand or sit 1-2 meter in front of the patient.
 - Ask the patient to follow your finger with their eyes without moving their head.
 - Check gaze in the six cardinal directions using a cross or "H" pattern.
- Pause during upward and lateral gaze to check for nystagmus.
- Check **convergence** by moving your finger toward the bridge of the patient's nose.
- Check **conjugate** movements.
 - By moving an interesting toy around in the child's field of view.
- Test Pupillary Reactions to Light (*association with CNII*)

Trochlear nerves (IV)

Ocular movements

Generalities

The smallest of the cranial nerves

Exclusively motor / innervates the Superior Oblique muscle.

Functions

Moves the eye downward and inward (medial)

Nerve Disorders



Isolated fourth n. palsy

- Not common.
- One of the important causes is head trauma in which the damage is sometimes bilateral.
- The eyeball is rotated outward by unopposed action of inferior rectus m.
- Diplopia only below the horizontal plane.

How to Examine

- Test Extra-ocular movements

Abducens nerves (VI)

Ocular movements



Functions

Exclusively motor / innervates the Lateral Rectus muscle which abducts the eye (moves it outwards / Lateral deviation).

Nerve Disorders

Isolated Nerve palsy.

- Common, because it has the longest intracranial course.
- Medial deviation of eyeball due to unopposed action of medial rectus m., diplopia on looking laterally.

- Causes: head injury ; increased intracranial pressure ; suppurative otitis media called Gradenigo's syndrome ; lesions of cavernous sinus (aneurysm, pituitary adenoma, meningioma)
- 6th nerve palsy + ipsilateral 5th and 7th nerves palsies + contralateral hemiplegia, occurs in unilateral pontine stroke.

Clinical Signs

Internal rotation of the eye / convergent paralytic squint of the affected side.

How to Examine

Test Extra-ocular movements.

Trigeminal nerves (V)

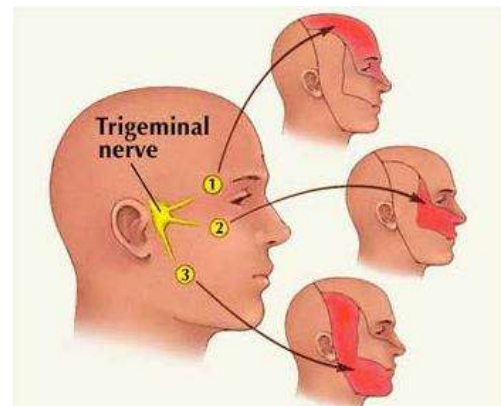
Somatic & Sensory

Generalities

Is the largest cranial nerve and the great sensory nerve of the head and face, and the motor nerve of the muscles of mastication.

2 roots, a small *motor* and a large *sensory*.

3 large divisions: the **ophthalmic**, **maxillary**, and **mandibular**. The ophthalmic and maxillary consist exclusively of sensory fibers; the mandibular is joined outside the cranium by the motor root.



Ophthalmic Nerve (1st division), is a sensory nerve. It supplies branches to the cornea, ciliary body, and iris; to the lacrimal gland and conjunctiva; to the part of the mucous membrane of the nasal cavity; and to the skin of the eyelids, eyebrow, forehead, and nose.

Maxillary Nerve (2nd division), is a sensory nerve. The side of the nose, the lower eyelid, and the upper lip, joining with filaments of the facial nerve.

Mandibular Nerve (3rd division) is a mixed nerve. Supplies the teeth and gums of the mandible, the skin of the temporal region, the auricula, the lower lip, the lower part of the face, and the muscles of mastication; it also supplies the mucous membrane of the anterior two-thirds of the tongue.

Temporal & Masseter muscles (jaw clenching) & lateral movements of the jaw are innervated by the Masseteric nerve of the mandibular division. Masseter is one of the most powerful muscles for its size in the body. It acts to raise the jaw and clench the teeth. This muscle functions to chew food, and is associated with angry and aggressive states.

Functions

- Ophthalmic division:
Forehead, corneal sensation & corneal reflex.
- Maxillary division: Infraorbital, palatal, & upper teeth sensation.
- Mandibular division:
Sensation: Mandibular, chin, lower teeth, & tongue.
Motor: Mastication, Tensor Tympani, Tensor Palati.

Nerve Disorders

Temporal-Mandibular Disorder

Clinical Signs

Neuralgia that is of very frequent occurrence

How to Examine

Test for:

1. Facial sensation

- In all 3 sensory divisions.
- Use the finger ("does this feel normal?") or a sharp stick ("does this feel sharp?")
- Ask the patient to report "sharp" or "dull"
- Compare sides if there are unilateral symptoms.

2. Corneal reflex (sensory)

Is dependent on the integrity of the 5th and the 7th cranial nerves.

- Distract the patient by asking them to gaze upwards.
- Lightly touch the cornea with a wisp of cotton wool, bringing the wool from the side of the eye.
- The corneal reflex should not be tested routinely, only if a disorder is suspected.



Interpretation of results

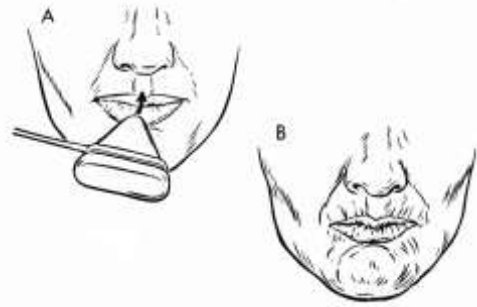
- Normal: both eyes should blink together with unilateral stimulation.
- Patient can feel the touch of the cotton wool (via the ophthalmic division of the trigeminal nerve), but there is no reflex blink (via facial nerve innervation of orbicularis oculi muscles). This indicates facial nerve palsy.
- Unable to feel touch of cotton wool, indicates trigeminal nerve palsy.

2. Examination of Masseter motor function

- Palpate the Masseter and Temporal muscles while:
- Clenching the teeth,
- Opening the mouth

3. Jaw jerk

- Can help to indicate an UMND
- Exaggerated in pseudobulbar palsy.



Procedure for testing of jaw jerk:

- The patient is told to let his mouth fall open slightly.
- The examiner places his finger on the patient's chin and taps his finger lightly with a tendon hammer.
- An exaggerated closure of the mouth can indicate an upper motor neuron lesion, e.g. pseudobulbar palsy of the 5th cranial nerve.

Facial nerves (VII)

Motor ; sensory ; special Sensory

Generalities

The facial nerve has 2 components. The larger portion comprises efferent fibers that stimulate the muscles of facial expression, scalp, and auricle. The smaller portion contains taste fibers to the anterior 2/3 of the tongue, secretomotor fibers (sympathetic motor fibers) to the lacrimal and salivary glands, and some pain fibers.

Functions

Facial expression.

Somatic sensory: posterior external ear canal.

Special sensory: taste (anterior 2/3 tongue).

Somatic motor: muscles of facial expression.

Visceral motor: salivary glands, lacrimal glands.

Clinical Signs

Motor:

Facial movements:

- Facial expression
- Closing the eye & the mouth

Sensory:

- Anterior 2/3 of the tongue / *Taste for salty, sweet, sour & bitter*



How to Examine

Always central VS peripheral / Focus attention on the voluntary movement of the upper part of the face on the affected side.

Motor Examination

- Observe for any facial droop, asymmetry in facial shape or in depth of furrows جعدة such as the nasolabial fold, lag, or weakness. Old photographs of the patient can often aid your recognition of subtle changes.
 - Test facial movement by asking the patient to do the following:
 - Raise eyebrows and frown (wrinkle the brow)
 - Shut both eyes tightly (to resistance)
 - Smile and show teeth
 - Corrugate the forehead (تجعيد الجبهة)
 - Purse lips يزوم الشفاه
 - Puff out (Inflate) cheeks
- 
- Patients with central (**upper motor neuron**) facial palsy:
 - Dorsolateral pons lesion due to superior cerebellar artery infarction.
 - Movements of the upper part of the face (brow wrinkling and, to a lesser extent, eye closure) are less affected than movements of the lower part.
 - Also, observe spontaneous facial movements (e.g., emotional smiling), since in some patients with central facial palsy, they will be better than movements to command.
 - **Explanation:**
In supranuclear lesions such as a cortical stroke (upper motor neuron; above the facial nucleus in the pons), the upper third of the face is spared while the lower two thirds are paralyzed. The orbicularis, frontalis, and corrugator muscles are innervated bilaterally, which explains the pattern of facial paralysis
 - Patients with peripheral (**lower motor neuron**) facial palsy:
Often have total hemifacial paralysis, and are unable to close the eye.

Sensory Examination

Assess the taste on the anterior 2/3 of the tongue with sugar, salt, or lemon juice on cotton swabs applied to the lateral aspect of each side of the tongue.

Acoustic nerves (VIII)

Special Sensory: Hearing ; Balance

Generalities

2 divisions:

1. Cochlear nerve (**Hearing**)
2. Vestibular nerve (equilibration / **Balance**)

Nerve Disorders

The following factors contribute to a high risk of deafness or hearing loss at birth:

- A family history of deafness.
- Hyperbilirubinemia.
- Congenital rubella syndrome.
- Defects of the ears, nose, or throat.
- Birth weight 1500 gm or less.

How to Examine

1. Hearing Examination

• **Finger Rubbing Test** (Screening)

- *Face the patient and hold out your arms with your fingers close to the patient's ears.*
- *Rub the fingers of one hand together while moving the fingers noiselessly on the other.*
- *Ask the patient to tell you when and on which side the sound of rubbing is heard. If light rubbing is not heard, increase intensity as needed and note any asymmetry.*
- *Both sides are compared.*

• **Audiometry** / Hearing levels:

- Normal hearing : 0-15 dB
- Slight: 15-25 dB
- Mild: 25-40 dB
- Moderate to severe; 40-55 dB
- Severe: 70-90 dB
- Profound: > 90 dB

2. Vestibular System Examination (Balance Testing)

Often vestibular dysfunction → nystagmus.

• **Barany test** (for positional vertigo).

- *The patient's head is rapidly lowered from vertical to about 30 degrees above the horizontal, and the examiner looks for nystagmus (rotary) over the next minute.*
- *The procedure is repeated with the head rotated to the left and then the right.*

• Ice-water Caloric Testing

- Use a minimum of 50 ml of ice water & position head at 30 degrees above the horizontal. Allow 1 minute after irrigating each ear and at least 5 minutes between testing on each side.
- Interpretation:
 - Absent Vestibulo-ocular reflex Testing:
 - No response to cold water stimulation = No deviation of the eyes, nor nystagmus.

Diagnosis of Neurofibromatosis Type 2 (Acoustic Neuroma)

- 1) Bilateral masses of the 8th cranial nerve seen with appropriate imaging techniques (CT or MRI).
- 2) A first-degree with NF2 and either:
 - i) Unilateral mass of the eighth cranial nerve,
or
 - ii) Two of the following:
 - (1) Neurofibroma
 - (2) Meningioma
 - (3) Glioma
 - (4) Schwannoma
 - (5) Juvenile posterior subcapsular lenticular opacity

The criteria are met by an individual who satisfies condition 1 or 2.

Glossopharyngeal nerves (IX)

Motor and sensory

Generalities

Contains both motor and sensory fibers, and is distributed, as its name implies, to the tongue and pharynx.

It is the nerve of ordinary sensation to the mucous membrane of the pharynx, fauces الحلق, and palatine tonsil, and the nerve of taste to the posterior part of the tongue.

Functions

- Somatic sensory and special Sensory (Afferent): taste and sensation of the posterior $\frac{1}{3}$ of the tongue, pharynx and middle ear (*posterior portions of the eardrum*).
- Visceral Sensory (Afferent): Carotid Body / Sinus.
- Somatic Motor (Efferent): Stylopharyngeus ; Pharynx.
- Visceral Motor (Efferent): Parotid gland.

IX, X. Glossopharyngeal & Vagus Nerves: Motor & sensory to the palate and vocal cords.

Clinical Signs

- Palatal movement paralysis (no elevation on saying Ah). The weak side of the palate fails to elevate or elevates weakly.
- Absence of Gag reflex.
- Vocal paralysis: Hoarse voice (Hoarseness) with difficulty opposing cords on saying "e".
- Loss of taste on the posterior $\frac{1}{3}$ of the tongue.
- **Progressive bulbar palsy:**
A form of motor neuron disease characterized by dysfunction of the muscles controlled by the cranial nerves of the lower brain stem (the "bulb") specifically, the Glossopharyngeal nerve (IX), Vagus nerve (X), and hypoglossal nerve, Manifested by weak palate, decreased gag, and nasal speech.
- **Pseudobulbar palsy:**
A form of motor neuron disease resulting from the degeneration of corticobulbar pathways to V, VII, X, XI and XII cranial nerve nuclei with sparing of the III, IV and VI nerve nuclei.
Is often associated with dysphagia, dysphonia and dysarthria, weak palate, exaggerated gag and nasal speech.

How to Examine

Examination is made for both Glossopharyngeal (IX) & Vagus (X) together

Sensory Examination of Glossopharyngeal (IX) & Vagus (X)

- Ask the patient to Say "Ah". Observing palatal movement / Palatal elevation (IX & X).
- Test the gag reflex with tongue depressor / *pharyngeal sensation (IX)*.
- Listen to the voice / is it hoarse / *Recurrent Laryngeal nerve dysfunction (X)*.

Sensory Examination of Glossopharyngeal (IX) & Vagus (X)

- Test the taste on the posterior $\frac{1}{3}$ of the tongue (IX)

Attention: Each side should be tested.

Vagus nerves (X)

Generalities

The Vagus has the most extensive distribution of cranial nerves, innervating the heart and the major part of the respiratory and alimentary tracts.

Has a mixed visceral afferent and efferent nucleus. It receives sensory fibers from the heart, the lower respiratory tract and the alimentary tract down to the transverse colon; in addition it gives rise to motor fibers to the heart and the smooth muscles of the bronchi and gut.

Functions

- Somatic sensory (afferent): external ear
- Visceral sensory (afferent): aortic arch/body
- Special sensory (afferent): taste over epiglottis
- Somatic motor (efferent): soft palate, pharynx, larynx (vocalization and swallowing).
- Visceral motor (efferent): broncho-constriction, peristalsis, bradycardia, and vomiting.

Pathology

1. Isolated lesions of the Vagus nerve are uncommon but it may be involved in injuries or disease of related structures.
2. Vagotomy
3. Injuries to the recurrent laryngeal nerve

How to Examine

Together with Glossopharyngeal (see above).

Spinal Accessory nerves (XI)

Motor

Generalities

Consists of 2 parts:

1. **Cranial** (is the smaller of the two).
2. **Spinal**.

Functions

Exclusively somatic motor (Efferent): Trapezius, Sternocleidomastoid

Pathology

1. Isolated lesions of the cranial root of the accessory nerve are infrequent; more commonly it is involved concomitantly with the Vagus when it gives rise to paresis of the laryngeal and pharyngeal muscles, resulting in dysphonia and dysphagia.
2. Division of the fibers of the spinal root (or lesions affecting their cells of origin) results in paresis of the Sternocleidomastoid and Trapezius muscles. This follows, for example, most block dissections of the lymph nodes of the neck, the nerve being sacrificed in clearing the posterior triangle.

Clinical Signs

- Dysphonia ; Dysphagia
- Paralysis / paresis of the Sternocleidomastoid (SCM) & upper portion of the Trapezius muscles.

How to Examine

Unilateral SCM Examination

- Rotate the head to the opposite side, then palpate and watch the SCM during active head turn to the opposite side against resistance.

Bilateral SCMs Examination

- Flex the neck forward against resistance.

Trapezius bulk examination

- Can be roughly assessed. Observe and palpate while the patient is shrugging (هز الكتفين إستهجائاً أو لا مبالاة) the shoulders against resistance.

Hypoglossal nerves (XII)

Motor

Generalities

- The hypoglossal nerve as the name indicates can be found below the tongue.
- The tongue is divided into lateral halves by a median fibrous septum which extends throughout its entire length and is fixed below to the hyoid bone.
- In either half there are two sets of muscles, extrinsic and intrinsic; the extrinsic muscles have their origins outside the tongue, the intrinsic are contained entirely within it.

Functions

Somatic Motor: Tongue

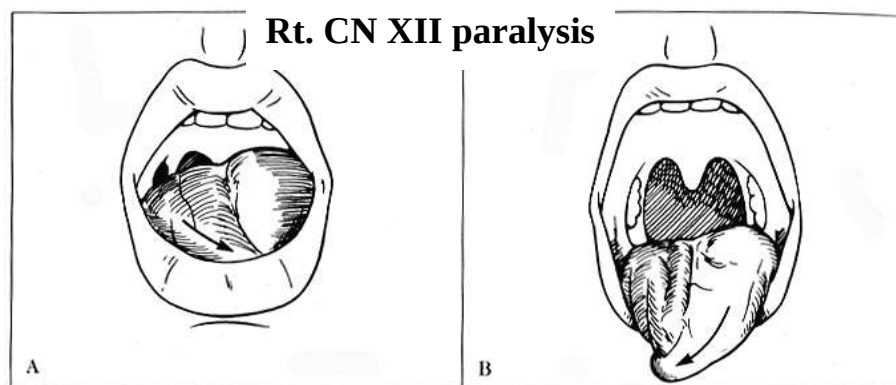
- This nerve is a somatomotor nerve (exclusively motor / efferent) that innervates all the intrinsic and all the extrinsic muscles of the tongue with the exception of the palatoglossus innervated by Vagus nerve. So all the tongue movements in any direction (sticking out, pulling forward, downward, backward and upward) are controlled by it.
- The movements produced by the extrinsic muscles are, however, somewhat coarse. Fine shape changes are assured by the intrinsic tongue muscles.
- By coordination of these 2 groups, this organ gets the power of assuming the forms necessary for the enunciation (لفظ) of the different consonantal sounds.

Nerve Disorders

- Tumor at the base of the skull, strokes, infections of the brain stem, or an injury to the neck, including that due to surgery, can damage the hypoglossal nerve producing its paralysis. Consequently, the tongue becomes weak on the affected side and eventually wastes away (atrophy).

Clinical Signs

- Fasciculations produced by muscular denervation (*Fasciculations without atrophy are often normal*).
- Tongue atrophy of the paralyzed side (*coving تجويف along the lateral surface*).
- Deviation of the tongues to one side:
 - When the tongue at rest / deviation to the opposite side.
 - When the tongue protruded / to the paralyzed side



B..... Tongue protruded

A..... Tongue at rest

- Difficulty in speaking (*can't assume the forms necessary for the enunciation of the different consonantal sounds*).
- Difficulty in chewing (*due to tongue movement deficits*)
- Difficulty in swallowing (*due to tongue movement deficits*).

How to Examine

- Observe the tongue as it lies in the mouth for:
 - Evaluation of the mass and contour of the tongue for possible tongue atrophy.
 - Deviation to one side or the other and its direction:
 - When at rest inside the mouth / ask the patient to open the mouth
 - When the tongue protruded / ask the patient to stick out the tongue, and
 - Fasciculations
- Listen to the articulation of the patient's words.
- Observe while chewing swallowing.

Meningeal Signs

Are the manifestation of meningeal irritation as:



- Headache
- Nuchal rigidity
- Back pain
- Kernig's sign
- Brudzinski sign

In some children, particularly in those younger than 12-18 mo, neck stiffness, Kernig and Brudzinski signs are not consistently present, so their absence could not rule out meningitis in children younger than 12 months.

1. Neck Stiffness

Stiffness of the neck resulting from infection and/or inflammation of the membranes covering the brain and spinal cord.

- An involuntary muscle spasm causing limitation of passive neck flexion.
- Put the child on his back (supine position)
- Place the hand on the child's occiput
- Attempt flexing the child's neck.
 - **Positive (neck stiffness is present):**
if lifting the whole trunk.
- The child him self is:
 - Unable (not unwilling) to flex the neck to place chin on chest.
 - Unable to kiss his knee.
 - Unable to touch his toes.
.....without bending his knees.



Medical conditions, other than meningitis, as possible causes of stiff neck as a symptom:

- Infections such as flu,
- cervical spine infection,
- physical neck disorders such as neck injury, neck strain (شد), neck sprain(التواء), whiplash (انحناء سريع),
- neck muscle disorders such as arthritis, disc disorder, torticollis, congenital cervical rib, poor sleeping posture, and wrong pillow.

2. Kernig's Sign

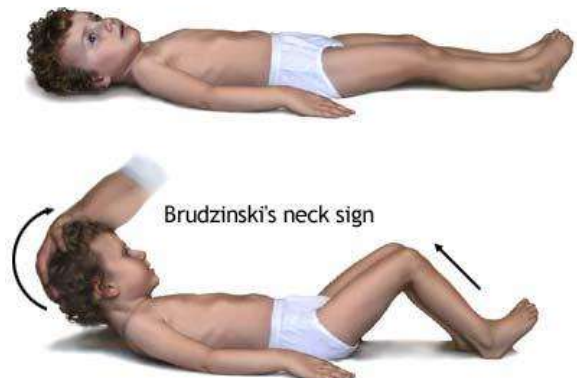
- Put the child on supine position.
- Move the hip and knee to 90°.
- Attempt at extending the knee fully.
- **Negative** (*Kernig's sign absent*):
If extension is easily achieved.
- **Positive** (*Kernig's sign present*):
 - If considerable resistance is faced (long before the leg is fully extended / can't be straightened).
 - If severe pain is felt in the hamstrings (becomes spastic) and the small of the back (flexion of the hip 90 degrees with subsequent pain with extension of the leg).



3. Brudzinski Sign

Is due to exudate around the roots in the lumbar region

- Put the child on supine position
- Flex the neck towards the chest
- **Negative** (*Brudzinski sign absent*):
 - If no involuntary flexion of the knees and hip is achieved.
- **Positive** (*Brudzinski sign present*):
 - When both knees and hips are involuntary flexed.



Motor Examination

Assess the integrity of the musculoskeletal system and the search for abnormal movements that may indicate an abnormality of the CNS or the peripheral nervous system.

Includes assessment of:

- **Muscular Power**
- **Muscle Bulk (trophicity)**
- **Muscular Tone**
- **Locomotion & Motility**
- **Reflexes (Primitive, Deep, Superficial)**

Motor Development

Neonate



- Upper & lower limbs in flexion.
- Head lag (when pulled in semi-sitting position).
- In ventral (prone) position, starts to move the head from one side to another.

Spontaneous Movements

The normal movements of newborns are jerky مرتج and usually alternate in the legs but are symmetrical in the arms. They may be jittery شديد العصبية or tremulous مرتعش. The limbs are usually flexed. Premature infants, on the other hand, show greater tendency to extension of the limbs, and their spontaneous movements may be writhing يتلوى and athetotic. Possible abnormalities include deviations from these characteristics, asymmetry, or abnormal movements such as myoclonus or convulsions.

Age 2 months

- Starts to maintain the head at the same axis of his back (at the prolongation of the back).
- In vertical position (in mother hands) can maintain the head, for a while, in vertical position.

Age 3 months

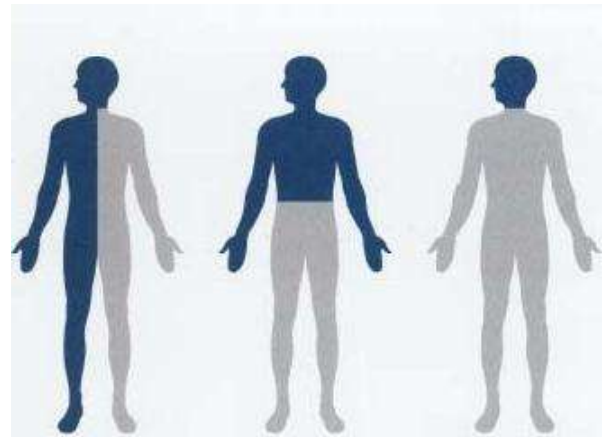
- Can assist by flexing the head in semi distance to sitting position.
- In sitting position: head could be maintained but the back is rounded.

Muscular Power (*strength*)

- The dominant side is slightly stronger (usually the right side).
- The muscle is stronger when shorter and weaker when longer.
- Extensors are less powerful on upper limbs in comparison with the lower ones, while flexors are more powerful on upper limbs in comparison with lower ones.

Categories of muscle weakness

- **Paresis:** Weakness
- **Plegia:** Paralysis / absence of strength
- **Hemiparesis:** Weakness of one half of the body.
- **Hemiplegia:** Paralysis of one half
- **Paraplegia:** Paralysis of the lower limbs.
- **Quadriplegia:** Paralysis of all 4 limbs
- **Diplegia:** Quadriplegia but with upper limbs less affected than the lower limbs.



Hemiplegia

Paraplegia

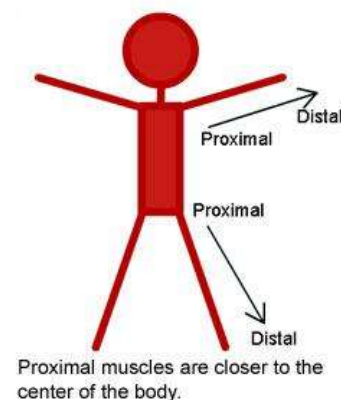
Quadriplegia

Muscle Strength Estimation Scale 0-5

Complete absence of muscular contraction / of course with no movements	0
Trace of contraction without displacement in space / no movements	1
Movements with gravity eliminated / only at the bed surface	2
Movements against gravity / above the bed surface	3
Movements against gravity with some resistance (but less than normal)	4
Movements against full resistance (normal muscle strength)	5

Assess:

- Muscle groups – segments – limbs.
- Compare between upper and lower limbs and between opposite limbs.
- Distal VS proximal.
- Flexors VS extensors.



Muscular Power Estimation

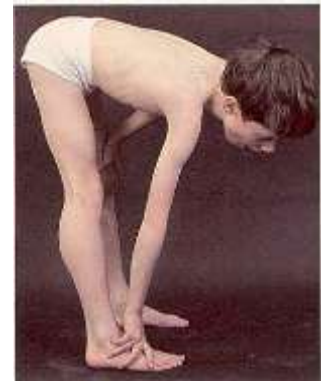
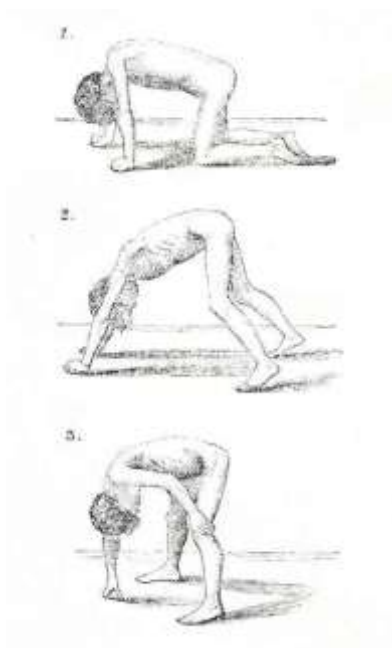
1. **Direct**-straight-forward in cooperative child / by request.

2. **Indirect-different ways** (small ages / non-cooperative):

- Squeeze the examiner's fingers.
- Passive movements:
 - Flex and extend the wrist and elbow.
 - Adduct and abduct the shoulder, the thighs against resistance.
- Shoulder girdle muscle strength / newborn or infant / supporting the child by the axilla / slips through if weakness.
- Palmar grasp /distal power.

3. **Observe:**

- Manipulation of objects.
- Climbing of stairs or stands from a sitting position: **Gower's sign**.



- Refusal to support body weight when suspended from axilla (diminished power in lower limbs).
- Decreased spontaneous activity of legs.
- Respiratory muscles (breathing):
 - Action of inter-costal muscles.
 - Diaphragmatic movements.
 - Use of accessory muscles.
 - Paradoxical breathing.

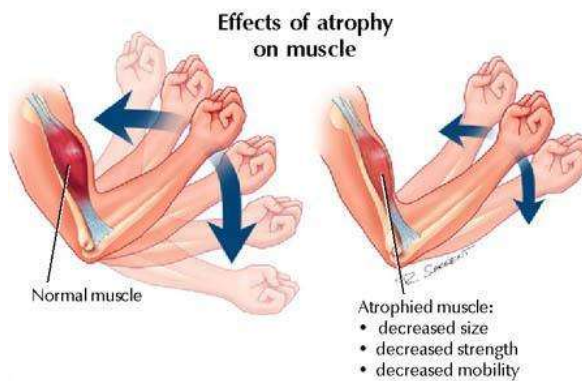


Muscle Bulk (trophicity)

Search for

Atrophy (wasting)

Shrinkage or wasting away of a tissue or organ because of a reduction in the size or number of its cells.



..... Global, distal or proximal.

Pseudohypertrophy

A term usually used to refer to the calf muscles in Duchenne muscular dystrophy, where the muscles appear hypertrophied while they are in fact weak.

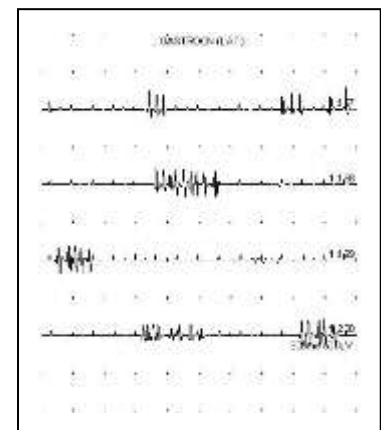
- The excess of body fat in most infants hides muscular atrophy.
- Atrophy and fasciculation in the tongue are sensitive clues.



Fasciculations

Are small, involuntary muscular contractions that are visible under the skin.

Fasciculation is a feature of lower motor neuron disease. Generally, it is **benign** if not associated with other signs of a motor lesion.



Muscular Tone

Tonus:

The slight residual tension of a normal muscle in the state of voluntary relaxation. (*The slight tension maintained by normal muscle even when it is relaxed*).

Tone disorders:

- Hypotonia / *Decreased tonus*
- Hypertonia / *Increased tonus*



Normal babies at birth (in a way or other) may be found to be "floppy" with head lag, trunk hypotonia and limbs with no resistance to passive motion. With time normal babies get out of it by the end of the neonatal period.



Examination of muscular tone:

- Ask the patient to relax.
- Flex and extend the patient's fingers, wrist, elbow, ankle and knee.
- There is normally a small, continuous resistance to passive movement.
- Observe for decreased (flaccid) or increased (rigid/spastic) tone.

Hypotonia



Hypotonia means decreased muscle tone. Infants with hypotonia seem floppy and feel like a "rag doll" *العبة شرائط* does when held.



Types:

- **Axial Hypotonia** with difficulty in maintaining head and spine (**head lag & spinal hypotonia**)
- **Peripheral Hypotonia** (of the extremities).
- **Generalized Hypotonia** / Floppy baby ; Hypotonic infant ; Axial and peripheral.

A newborn baby has no control over his head when pulled to a sitting position. By 8 wk. a baby has some muscular control and head lag is seen to be less than it was in early weeks. It is not until 4 months that a baby is expected to show no signs of head lag.

Neonatal generalized hypotonia is often a sign of a worrisome abnormality and may suggest the presence of CNS dysfunction, metabolic, genetic, or muscle disorders.



Axial hypotonia is reflected in head lag when pulling the child to sitting from lying down. Head control may be poor or absent in the floppy infant with the head falling to the side, backward, or forward, with overextension of the back. In sitting position there is overflexion of the back.

In **generalized hypotonia** poor sucking and chewing may also be present in some hypotonic children depending on the underlying condition. Their elbows and knees are loosely extended, while infants with normal tone tend to have flexed elbows and knees.

Infants with normal tone can be lifted with the parent's hands placed under the armpits, while hypotonic infants tend to slip between the hands as the infant's arms rise unresistingly upward.

Hypotonia can be a symptom of many different disorders. Its severity can vary significantly from mild to severe hypotonia depending on the underlying cause. Some children may end up with serious conditions, some with mild conditions and some will end up normal.

Hypotonia is not the same as muscle weakness but it can co-exist with muscle weakness.

Hypertonia

- 2 Types:
 1. Spasticity
 2. Rigidity

Spasticity

Increased tone with initial resistance to passive movements, followed by a sudden release called the “**clasp knife**”. Worse at the extremes.



- Most apparent in:
 - The upper extremity flexors.
 - Lower extremity extensors.
- Could be associated with brisk tendon reflexes, clonus, diminished active movements and disuse atrophy.
- It is a manifestation of an upper motor neuron lesion.
- Could be bilateral or unilateral.
- Child with lower limb spasticity drags the legs while crawling and walks on tip toes and drags the spastic limb.



Physiopathology of spasticity

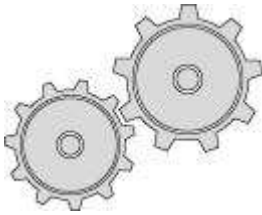
- In the case of spinal cord injury, the nerve cells below the level of injury become disconnected from the brain at the level of injury with scar tissue formation, blocking messages between areas below the level of injury and the brain.
- Spasticity does not occur immediately. The spinal cord

goes first into spinal shock (changes to the nerve cells which control muscle activity) expressed by flaccidity and hypo or areflexia. This may last up to several weeks.

- Once spinal shock wears off, the natural reflex which is present in everyone reappears.
- Spasticity is an exaggeration of the normal reflexes that occur when the body is stimulated in certain ways. In normals, a sensory signal is sent through the reflex arch where it travels to the brain via the spinal cord. The brain then assesses the stimulant, and if the stimulant is thought not to be dangerous, an inhibitory signal is sent down the spinal cord, and cancels the reflex from moving the muscle. In a person with a spinal cord injury this inhibitory signal is blocked by the structural damage in the cord, and the natural reflex is allowed to continue resulting in a contraction of the muscle.
- Once spasticity is established, the chronically shortened muscle may develop physical changes such as shortening and contracture that further contribute to muscle stiffness.
- The best way to manage or reduce excessive spasms is to perform a daily range of motion exercise program (physiotherapy).
- Spasticity usually is accompanied by paresis and other signs, such as increased stretch reflexes, collectively called the upper motor neuron syndrome. Paresis particularly affects distal muscles, with loss of the ability to perform fractionated movements of the digits.

- The upper motor neuron syndrome results from damage to descending motor pathways at cortical, brainstem, or spinal cord levels.
- The interval between injury and the appearance of spasticity varies from days to months according to the level of the lesion.
- In addition to weakness and increased muscle tone, the signs in spasticity include clonus, the clasp-knife phenomenon, hyper-reflexia, and the Babinski sign.

Rigidity



A constant resistance to passive movements of both extensors and flexors. “**Lead pipe**” persists through out the range and in both directions.

- Results from basal ganglia lesion.
- Persists with repetitive passive extension and flexion of a joint and doesn’t give away such as spasticity.
- A typical “**cogwheel**” [دولاب مسنن] sensation may be evident.



Generalized hypertonia (*spastic or rigid*)

- The lower limbs are usually more affected than the upper limbs.
- The trunk tends to be opisthotonic.
- The extensor tone in the neck results in seemingly good head control in ventral suspension but poor head control on pulling to sit.

Examination of Coordination

To produce a smooth, accurate and purposeful movement, a coordinated combination of a series of motor actions is needed. This requires 4 CNS components in an integrated way:

- Motor System (*muscle strength*).
- Cerebellar System (*rhythmic movements and steady posture*).
- Vestibular System (*balance and coordinating eye, head and body movements*).
- Sensory System (*position sense*).

Ataxia

Abnormalities of movement characterized by decomposition of movement.

Complex movements are broken down into component parts, so that movements appear jerky and confused.

1. Primarily Truncal Ataxia

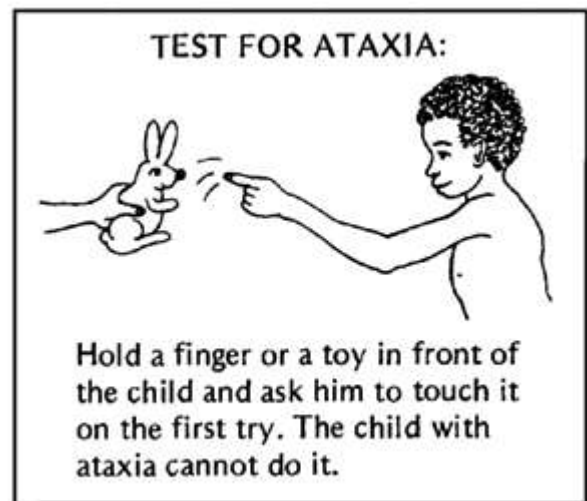
- Truncal unsteadiness during sitting or standing (inability to maintain truncal posture, with variations in any direction that can become rapidly rhythmic ("titubation").
- In severe cases, this may affect the neck and trunk even when patients are seated (static ataxia).
- Ataxia of axial (truncal) and leg muscles (dynamic ataxia) leads to a broad-based unsteady gait (ataxic gait / wide-based unsteady gait).
- The patient may fall in any direction.

Cause: cerebellar vermis pathology.

2. Extremities Ataxia

- Kinetic tremor (*Intentional*)
- Seen with limb movements.
- Unaffected by visual attention.

Cause: cerebellar hemispheres pathology.



3. Sensory Ataxia

- Unsteadiness (positive Romberg sign) with eyes closed / not opened.
- Often associated with sensory findings (abnormal joint position and vibration sense).

Cause: spinal cord and peripheral nerve disorders.

Dysdiadochokinesia

- Decomposition of distal movements reflected in diminished performance or inability to perform rapid alternating movements.
- From Greek dys "bad", dia "across", docho "receive", kinesia "movement".
- Movements can't be followed quickly by its opposite and are slow, irregular and clumsy.

Rapid Alternating movements Test

- Ask the patient to repeatedly strike one hand on the thigh, raise the hand, turn it over, and then strike it back down as fast as possible.
- Ask the patient to repeatedly tap the distal thumb with the tip of the index finger as fast as possible.
- Ask the patient to tap your hand with the ball of each foot as fast as possible.
- Mainly in cerebellar disease. Also in upper motor neuron & basal ganglia disorders. The non-dominant side often performs less well.



Dysmetria

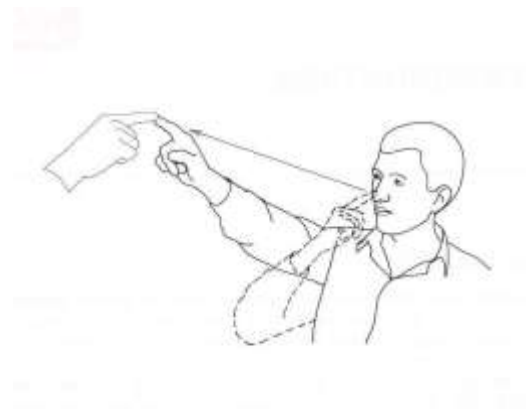
Error in measuring distances.

- The trajectory of movements is disturbed, so that movements may be:
 - Too small (**Hypometria**)
 - Too large (**Hypermetria**)
 - May miss their target.

1. Point to Point Movements Tests

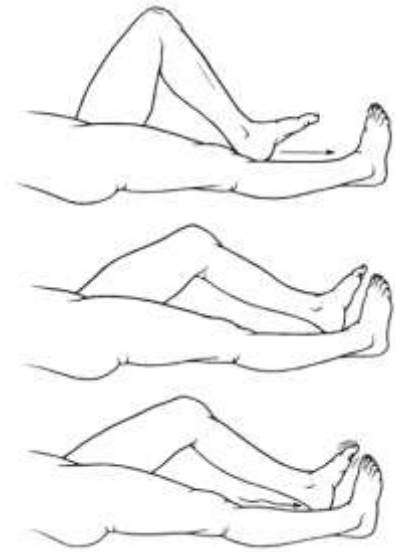
• **Finger-to-Nose** (*Test for both sides*)

- Ask the patient to touch your index finger and their nose alternately several times with eyes opened then closed.
- Move your finger about as the patient performs this task.



• Heel-to-Shin Test

- Ask the patient to place one heel on the opposite knee and run it down the shin to the big toe.
- 1st with eyes opened then closed.
- Test for both sides.
- Observe the accuracy, smoothness and for tremor.



Findings:

- In cerebellar disease:
 - They may miss the target (Hypermetria) but finally reaches the mark.
 - Movements are clumsy, unsteady and inappropriately varying in their speed, force and direction. Are jerky in uncoordinated fashion (decomposition of movement).
 - Proximal tremor (fulcrum at the shoulder) that results in a tremor perpendicular to the line of movement (side-to-side, perpendicular to the line nose-finger).
 - With eyes closed incoordination get worse.
- In hypothyroidism and “essential tremor”:
 - Terminal tremor (intentional).



2. Abnormal check & rebound ارتداد

- Rebound: inability to inhibit a muscular action reflected in impaired ability to stop movements abruptly.

How to examination:

- Sudden release of a flexed arm by the examiner and the patient inadvertently غير متعمد strikes the face.

Manifestations of Cerebellar Disease (*clinical signs*)

- Ataxia & dysmetria (*due to incoordination of axial* & appendicular muscles**).
- Dysarthria (due to incoordination of speech muscles).
- Nystagmus (due to incoordination of eye movements).
- Hypotonia (physiological basis unclear)
- Decreased DTRs

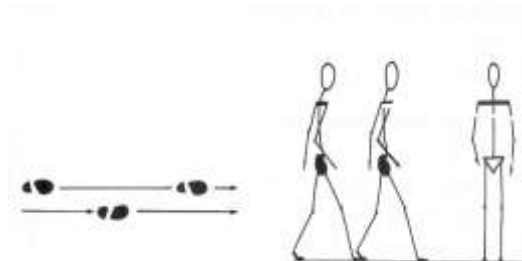


Examination of Gait

At the age of 2½-yr. the gait is steady, well coordinated and no longer wide-based. The arms are down to the side and there are normal associated movements. The hands are no longer used when getting up off the floor or sitting down on the floor. At this age, the child is able to run. By the age of 8-10 yrs. he will acquire the adult gait pattern.

Ask the patient to:

- *Walk across the room, turn and come back.*
- *Walk heel-to-toe in a straight line.*
- *Walk on their toes in a straight line.*
- *Walk on their heels in a straight line.*
- *Hop in place on each foot.*
- *Do a shallow knee bend.*
- *Rise from a sitting position.*



Normal Gait

Technique of examination

- *Stand still with the feet together (if possible).*
- *With eyes open and then closed.*
- *With eyes open to walk about ten paces, turn around, and walk back.*
- *Hands should be at the sides, and not carrying anything.*
- *If unstable, be sure to prevent falling down.*

* The axial muscles arise on the axial skeleton. They position the head and spinal column and also move the rib cage, assisting in the movements that make breathing possible. They do not play a role in movement or support of either the pectoral or pelvic girdle or the limbs. This category encompasses roughly 60 percent of the skeletal muscles in the body.

* The appendicular muscles stabilize or move components of the appendicular skeleton and include the remaining 40 percent of all skeletal muscles.

Observation of Gait

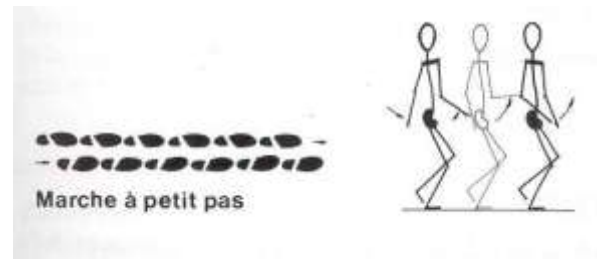
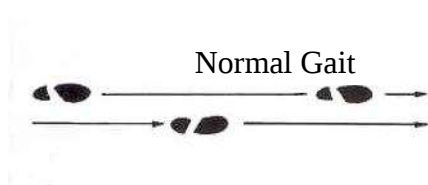


- Posture & stability (with eyes open and closed).
- Wideness of base (how far apart the patient must place the feet to feel steady.)
- Stride (pace) خطوة المشي (length, balance, & rhythm).
- Efficiency of turning.

Types of Gait

1. Short Pace Gait (*Marche a petits pas*)

- With Marked arm swing.



2. Spastic Gait (*scissoring gait*)

- Legs appear stiff.
- Thighs are tightly adducted, leading to crossing over.
- Dragged toes (tip toeing).

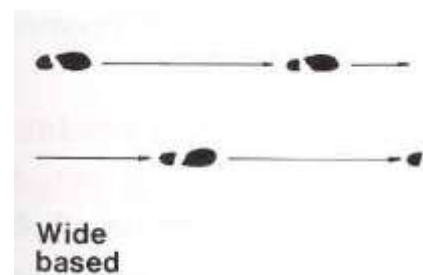
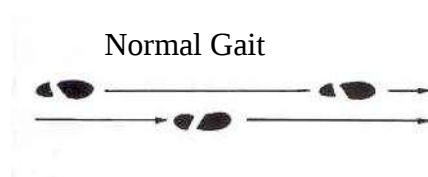
Cause: Bilateral Upper Motor Neuron Disease.



The legs stiffen and the feet go into a rigid tiptoe position.

This child is *not* almost ready to walk.

3. Wide-based gait



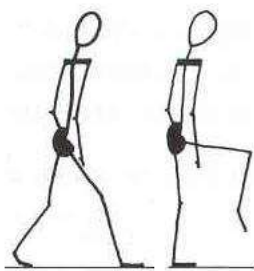
4. Cerebellar Ataxic Gait

- Widened base (broad-based)
- Irregular gait (uneven rhythm, unequal size of steps, variable direction).
- Closing the eyes makes the situation worse.



To keep her balance the child with ataxia walks bent forward with feet wide apart. She takes irregular steps, like a sailor on a rough sea or someone who is drunk.

5. Drop Gait (High Stepping Gait)



High stepping

- Knees lifted high.
- Patients compensate for foot drop (dorsiflexion absent or weak at ankle) by lifting the leg higher (flexing at the hip and knees) to prevent the toes from hitting the floor.

6. Waddling gait (مشي البطّة)



Syn. Proximal Muscle Weakness Gait ; Trendellenberg gait

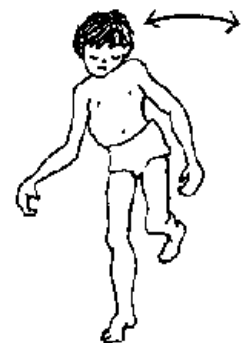
- A style of walking seen in a patient with proximal Myopathy, CHD and pregnancy.

It is characterized by:

- A broad-based gait with a duck-like

waddle . يتهدّى

- Marked rotation of pelvis and shoulder ; the pelvis drops to the side of the leg being raised.
- Forward curvature of the lumbar spine.
- Marked body swing



7. Hemiplegic Gait

- Asymmetrical.
- The affected leg swing out to the side.



8. Antalgic Gait

- Mostly asymmetrical.
- Associated with pain and/or with limitation and/or modification of the range of paces.

9. Orthopedic Gait

- Mostly asymmetrical
- A result of bony or joint deformity.

Reflexes

Primitive and non-primitive

Primitive Reflexes

- They develop during uterine life, in the 1st few weeks of fetal development. They should be fully present at birth and are gradually inhibited by higher centers in the brain during the first 6 to 12 months of post-natal life. As they are inhibited, another group of reflexes, called the **postural reflexes**, should emerge to help the infant or child to cope with demands of a gravity-based environment, and provide the basis for the control of automatic balance, posture and voluntary movement. The postural reflexes are regulated by higher centers in the brain that are involved in the execution of voluntary movement.
- Provide an indication of the status of the CNS. If they are activated by minor stimuli in the environment at a later age, they can interfere with the development of more complex skills.
- Are automatic survival stereotyped patterns, as ingrained (متأصل) in our behavior as the 'fight or flight' response with which we are so familiar. Unlike fight/flight responses, primitive reflexes are to help us survive in the very vulnerable pre-walking stage of our life, when we are highly dependent on others for our needs.
- They are directed by a very primitive part of the brain (brain stem), executed without involvement of higher levels of the brain (the cortex). Ideally they are short lived and as each fulfils its function, it will be **replaced** by more sophisticated structures controlled by the cortex.
- **Primitive reflexes** (such as the Moro, grasp, and Galant) are normally present in the term infant and diminish over the next 4 to 6 months of life.
- The **postural reflexes** (such as the Landau and parachute) emerge at 3 to 8 months of age. Persistence of primitive reflexes and the lack of development of the postural reflexes are the hallmark of an upper motor neuron abnormality in the infant.
- Postural reflexes once present they should remain for life. Their absence is an indication that the CNS is immature.
- In some individuals, full integration and transformation of the primitive reflexes fails to occur and they remain active despite normal development in other areas. When this occurs, it contributes to the underdevelopment of efficient proprioceptive-motor integration, hand-eye co-ordination, lateral integration and aspects of perceptual performance.

- Detection of primitive reflexes (diagnostic assessment) can help isolate some of the causes of a child's problems so that remedial training can be targeted more effectively

The most important primitive reflexes are:

Rooting Reflex



A reflex seen in normal full-term newborn babies, who automatically turn their face toward the stimulus and make sucking motions with the mouth (rooting / roots for milk) when the cheek or lip or corner of the mouth is lightly touched. The baby lowers the bottom lip on the same side and his tongue moves towards the point of stimulation and turns his mouth towards the point of contact. The rooting

reflex helps to ensure breastfeeding.

Emerges at 24-28 weeks in utero. Present at birth and disappear by 9 months of age.

- Its absence at birth suggests general neurological depression, hypotonia, immaturity, bulbar palsy.



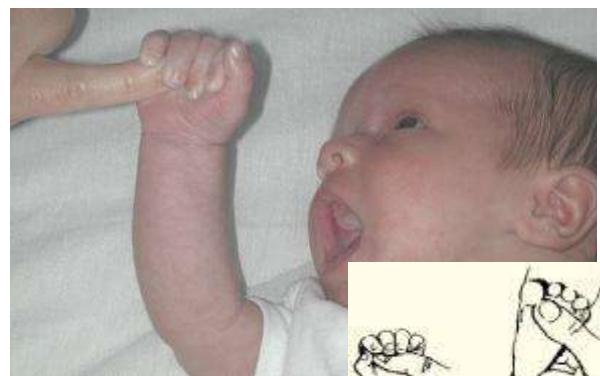
- If retained may affect swallowing, feeding, speech, articulation and manual dexterity in an older child



Palmar / Foot Grasp (reflex)

Place the examiner's finger in the infant's palm / sole. The hand closes round the examiner's finger (maintaining a grasp).

- Are fully present at birth and usually inhibited by 2-3 months of life for palmar and 7-8 months for plantar.



- The grasp reflex is replaced by the pincer grip قبضة فكي الكماشة at 36 weeks of age.

- Absence of this reflex suggests general neurological depression ; hypotonia ; hemi-syndromes / Erb's palsy ; clavicular fracture.
- It persists beyond normal time in spastic CP.

Asymmetrical Tonic Neck Reflex (ATNR)



Is activated as a result of turning the head to one side when the baby is lying on his back (supine). As the head is turned, the arm and leg on the same side will extend while the opposite limbs bend.

- Should be inhibited by 6 months of age.
- Its persistence at a later age is potentially a very handicapping disability, affecting seeing both hands simultaneously, mouthing, poor handwriting and severe restriction on the ability to achieve standing and walking.
- Is persistent in severe cerebral palsy.

Trunk Incurvation (Spinal Galant) Reflex



The trunk and hips move towards the side of the stimulus.

- Is present at birth and should be inhibited between 3 and 6- 9 months of post-natal life.
- If it persists it can affect ability to sit still, inattention, poor posture. Can contribute to the development of scoliosis of the spine. Sometimes associated with poor bladder control and bedwetting. May be retained on one side and so affects posture, gait and other forms of locomotion.

Parachute Reflex



The infant is held in vertical suspension and suddenly lowered toward a flat surface. The normal positive response is a forward extension of both arms and dorsiflexion of the infant's hands during the movement.

- This is a protective response that protects the infant if he falls.
- Appears before the onset of walking at about 5-6 months.

The parachute is the last of the postural reflexes to develop. It usually appears at 8 to 9 months and certainly is present by 12 months of age. The reflex is elicited by turning the child upside down. The arms should come forward and the hands spread out to catch the fall. Asymmetry of the reflex is abnormal and may indicate paresis in the non-extended extremity.

Moro Reflex

Synonym / Startle reflex



Is usually triggered if the baby is startled by a loud noise or if his head falls backward or quickly changes position.

- Present reflex will include spreading the arms and legs out widely with extension of the neck. Then quickly brings the arms back together and cry.
 - Is the earliest form of "fight or flight" (reaction to stress) which is fully present at birth. It disappears by 3-6 months.
- Sometimes there is a slight tremor or even a rhythmic shaking of the limbs.
 - Asymmetry usually suggests fracture of the clavicle or Humerus, injury to the brachial plexus (Erb's palsy), or neonatal hemiplegia.

Stepping / Walking reflex



- Most parents are surprised by this reflex.
- Held the baby under his arms, support his head, and allow his feet to touch a flat surface. Normal baby will appear to take steps and walk.
- Usually disappears by 2-3 months, until it reappears when the child learns to walk at around 10-15 months.

Placing Reflex

Flexion followed by extension of the lower limb when the infant is held erect and the dorsum of the foot is drawn along the under edge of a table top, seen in the normal infant up to the age of six weeks.



Landau Reflex



All normal neonates display some evidence of tone when suspended in the prone position. As the newborn infant is turned to prone, with the trunk or abdomen supported, the legs should be flexed.

- While holding the infant in ventral suspension with the head, spine, and legs extended, the nurse then passively flexes the head forward. The reflex is considered present if the whole body then flexes.
- The reflex may be seen as early as 3 to 4 months but should be present after 7 months of age.

Neither the **Landau** nor the **Asymmetrical Tonic Neck Reflex** are true primitive or postural reflexes, since they are not present at birth, and do not remain present for the remainder of life.

Non Primitive Reflexes

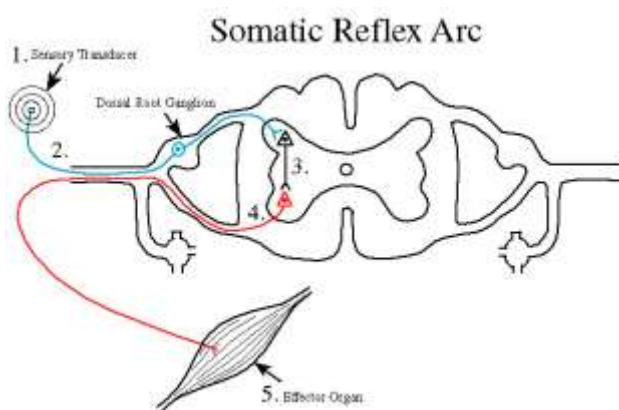
Reflexes can be categorized as either autonomic or somatic.

Autonomic reflexes

Are not subject to conscious control, are mediated by the autonomic division of the nervous system, and usually involve the activation of smooth muscle, cardiac muscle, and glands.

Somatic reflexes

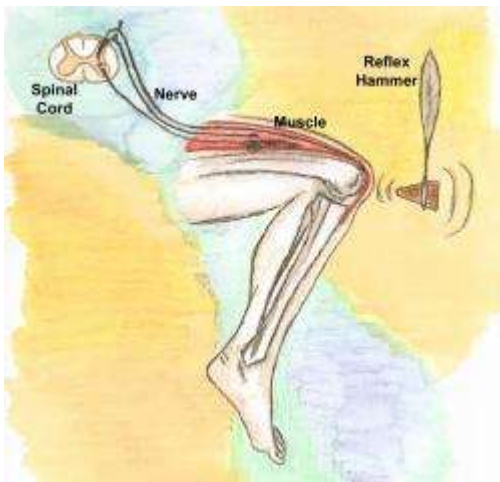
Involve stimulation of skeletal muscles by the somatic division of the nervous system.



Reflex arcs have five essential components:

1. The receptors at the end of a sensory neuron (react to a stimulus).
2. The sensory neurons (conduct nerve impulses along an afferent pathway towards the CNS).
3. The integration centers (consist of one or more synapses in the CNS).
4. Motor neurons (conduct a nerve impulse along an efferent pathway from the integration center to an effector).
5. Effectors (respond to the efferent impulses by contracting (If the effector is a muscle fiber) or secreting a product (if the effector is a gland)).

The importance of reflex testing



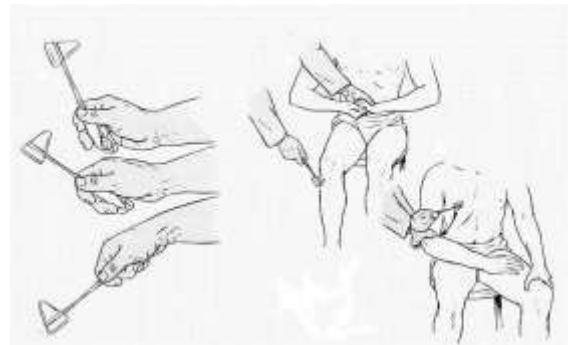
- Reflex testing is an important diagnostic tool for assessing the condition of the nervous system.
- Decreased, exaggerated, or absent reflex responses may indicate degeneration or pathology of portions of the nervous system, often before other signs are apparent.
- If the spinal cord is damaged, then reflex tests can help determine the area of injury. For example, motor nerves above an injured area may be

unaffected, whereas motor nerves at or below the damaged area may be unable to perform the usual reflex activities.

- Closed head injuries, such as bleeding in or around the brain, may be diagnosed by reflex testing. Remember that the Oculomotor nerve stimulates the muscles in and around the eyes. If pressure increases in the cranium (such as from an increase in blood volume due to brain bleeding), then the pressure exerted on CN III may cause variations in the eye reflex responses.

Technical aspects

- Briskly tap the tendon of a partially stretched muscle.
- One of the ways to reduce apprehension in the child is to place your thumb or index finger over the tendon to be examined and tap on it rather than directly tapping on the child's tendon. It's less threatening.
- Position the patient properly before starting.
- Reflex response depends on the force of your stimulus. Use no more force than you need to provoke a definite response.



- Reflexes can be reinforced by having the patient perform isometric contraction of other muscles (clenched teeth) or **hooks together his flexed fingers and pull apart (Jendrassik maneuver)**.
- In infants, the ankle DTR is difficult to obtain by tapping the Achilles tendon. Instead: gently dorsiflex the foot and tap the plantar surface. This gives a response.
- The biceps, knee and ankle DTRs / the most reliable in premature and infant.

Tendon Reflex Grading Scale

Areflexia	Absent no response	0
Hyporeflexia	Only elicited with reinforcement	0.5 +
Hyporeflexia	Diminished.	1+
Normal		2+
Hyperreflexia	More reflexive than normal without clonus	3+
Hyperreflexia	Very brisk with clonus	4+
Hyperreflexia	Sustained clonus	5+

DTRs Abnormalities

• Absent or Decreased

- Myopathies
- Peripheral Neuropathies
- Neuromuscular Junction Disorders
- Cerebellar abnormalities

• Increased

- Upper motor neuron lesions

Clonus Jerkiness (Clonus)

- If the reflexes are hyperactive, test for ankle clonus by sudden dorsiflexion of the foot with the knee partially flexed.
- Observe for **rhythmic oscillations**.
- Sustained clonus is always abnormal.
- Up to about 5 beats in a newborn is a normal finding unless the clonus is asymmetrical.

Plantar Response (Babinski reflex)

(Extensor plantar reflex)

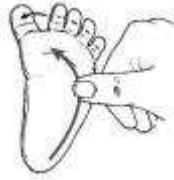
- Occurs when the great toe dorsiflex toward the top of the foot and the other toes fan out after the sole of the foot has been firmly stroked.
- One of the infantile reflexes.
- It is normally up in children under 2 years old.
- It disappears as the child ages and the nervous system becomes more developed.
- Over 2 years of age, the presence of a Babinski reflex indicates damage to the nerve paths connecting the spinal cord and the brain (the corticospinal tract).



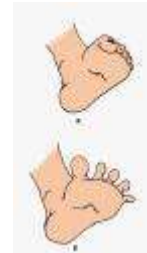
- Because this tract is right-sided and left-sided, a Babinski reflex can occur on one side or on both sides.
- An abnormal Babinski reflex can be temporary or permanent.

How to examine

- Stroke the lateral aspect of the sole of each foot with the end of a reflex hammer or key.



- Note movement of the toes, normally flexion (withdrawal). This is referred to as absent Babinski or Babinski down.
- Extension of the big toe with fanning of the other toes is abnormal. This is referred to as a positive Babinski.



Too vigorous stimulation may produce withdrawal, which may be misinterpreted as a Babinski response

Movements

normal and abnormal

How to describe a movement

Observe quality of:

- Gross motor movements / *trunk and limbs*
- Fine motor movements / *face, fingers and feet.*

These movements could be

..... *normal, hypoactive (paucity of movements) or hyperactive.*

- Asymmetrical movements.

Tics

Identical repeated movement (habit) عادة



- Is a habit in which a part of the body moves repeatedly, quickly, suddenly, uncontrollably and briefly. They are repeated in the same pattern for the same person. They consist of muscle (or a group of muscles) contractions.
- Tics can occur in any body part, such as the face, shoulders, hands or legs.
- The affected person is conscious and knowledgeable of presenting them and so becomes shy of having them.
- They can be stopped voluntarily for brief periods but not to a degree to control them fully.
- They disappear during sleep, but are exaggerated and became more frequent with anxiety and intense emotions.

- More frequent in the 5-7 yr. age group.
- 2 types:
 1. Motor tics expressed by movements
 2. Vocal tics expressed by sounds that are made involuntarily (such as throat clearing).
- Most tics are mild and hardly noticeable. However, in some cases they are frequent and severe, and can affect many areas of a child's life.
- The most common tic disorder is "**transient tic disorder**" or "**simple tic**". May affect up to 10% of children during the early school years.
- Some tics do not go away. Tics which last one year or more are called "chronic tics." or "complex tics". Chronic tics affect < 1% of children and may be related to a special, more unusual tic disorder called Tourette's Disorder (Gilles de la Tourette). Sometimes people with Tourette's Disorder may blurt out obscene (فاحش قذر) words (coprolalia / *The uncontrolled, often obsessive use of obscene or scatological language that may accompany certain mental disorders, such as Tourette's syndrome*), insult others, or make obscene gestures or movements.

Tremor



Is an involuntary, rhythmic oscillation of reciprocally innervated, antagonistic muscle groups, causing movement of a body part about a fixed plane in space.

May occur at rest or only on reaching for an object (intentional tremor of cerebellar disease).

- Frequency can be divided into three categories of oscillations per second:
 1. Slow (3 to 5 Hz)
 2. Intermediate (5 to 8 Hz)
 3. Rapid (9 to 12 Hz)
- Depending on the displacement produced by the tremor about the fixed plane (Amplitude) a tremor may be classified as:
 1. Fine
 2. Medium
 3. Coarse

A coarse tremor has a large displacement, whereas a fine tremor is barely noticeable.



May affect the head, face, jaw, tongue, trunk, extremities or voice.

On the basis of when it occurs, tremor is classified in:

1. Postural tremor

With a certain posture, is observed when the patient tries to maintain a posture against gravity, such as holding the arms out in front of the body.

2. Resting tremor

At rest it occurs when the patient is attempting to maintain the position of a body part at rest (e.g., when the patient's hands exhibit a tremor as they are resting in the patient's lap)

3. Action tremor (kinetic or intention tremor)

During action it occurs during movement of the affected body part from one point to another. A task-specific tremor occurs only when the patient begins to perform a highly skilled activity, such as writing or speaking.

- Tremor may be either:

1. Physiologic tremor

Is a normal variant, occurring at a frequency of 8 to 12 Hz in the hands yet as slow as 6.5 Hz in other body parts during maintenance of a posture. It can be increased by emotions such as anxiety, stress or fear, by exercise and fatigue, hypoglycemia, hypothermia, hyperthyroidism and alcohol withdrawal. When such an increase occurs, physiologic tremor is then called enhanced or exaggerated physiologic tremor. Certain drugs can also exacerbate physiologic tremor

2. Pathologic tremor

Is either idiopathic or occurs secondary to some disorders. Essential tremor and Parkinsonian tremor are two common types of pathologic tremor.

New method for extremity tremor detection and quantification based on optical measurements of tremor, using one or more cameras. Using video image processing, we are able to track any kind of extremity movements in real time (Tremor Tracker)

Titubation / head titubation

Tremor of the head and neck

It means also staggering (مربك) or unsteadiness of walk or posture.

It comes directly from the Latin verb titubare, which means “stutter” as well as “stumble” يتلعثم.

A family of five patients with Spinocerebellar Ataxia type 2 was described, This diagnosis was confirmed by molecular analysis. Patient 1, a 49-year-old man with titubation, dizziness, and loss of balance from the age of 34 years, had deteriorating speech and handwriting. Cerebellar signs included moderate gait ataxia, intention tremor, and dysdiadochokinesis, as well as titubation. Deep-tendon reflexes were all brisk.

Chorea



Rapid, involuntary uncontrolled, brief repetitive ceaseless, irregular, jerky, large-scale, dance-like movements.

Usually starts in one part of the body and moves abruptly, unpredictably, and often continuously to another extremity (major joints), trunk or face, which may interfere with speech or gait. The child is incapable of extending the arms without producing these abnormal movements.

They increase with effort or excitement.

Very rare. Sydenham chorea

Athetosis/oid



A continuous □stream of slow, involuntary, sinuous writhing يتلوى movements, usually of the proximal limbs (arms and hands).

The commonest cause is cerebral palsy but basal ganglia disease, in particular Wilson's disease, must be excluded.

Typical athetoid arm and hand movements may be as a regular shake or as sudden 'spasms'. Uncontrolled movements are often worse when the child is excited or tries to do something.



Choreo-athetosis

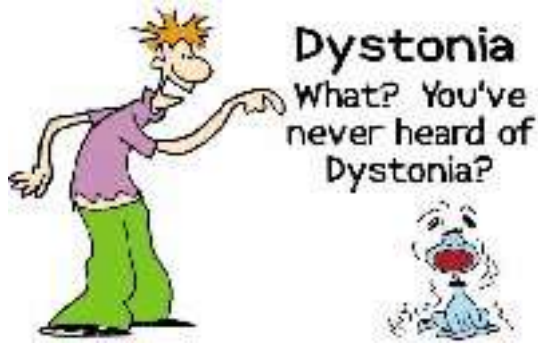
Chorea and athetosis, that occur together.

Are not disorders. Rather they are symptoms that can result from several very different disorders. Chorea and athetosis result from abnormalities in the basal ganglia, the part of the brain that helps smooth out and coordinate movements initiated by nerve impulses from the brain. In most forms of chorea, an excess of dopamine, the main neurotransmitter used in the basal ganglia, prevents the basal ganglia from functioning normally. Drugs and disorders that increase dopamine levels or increase the sensitivity of nerve cells to dopamine tend to worsen chorea and athetosis.

Chorea and athetosis occur in Huntington's disease, a hereditary disease.

Chorea may also be caused as Sydenham's chorea (a complication of rheumatic fever) that can last for several months.

Dystonia



Impairment of muscular tonus. Involuntary irregular, clonic contractions of the muscles of the trunk and extremities which twist the body forward and sideways

It is a sort of persistent abnormal posturing induced by muscular

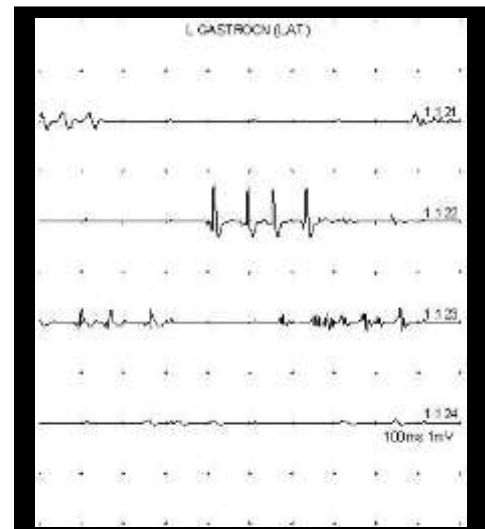


contraction.

Fasciculations

Are visible twitches (*visible under the skin*) in a resting muscle that usually is too small to result in joint movement (*tongue in Werdnig Hoffmann Disease / Spinal Muscular Atrophy*).

Fasciculation is a feature of lower motor neuron disease. Generally, it is benign if not associated with other signs of a motor lesion.



Epileptic & Non Epileptic Clinical Seizures

- Convulsive or non-convulsive.
- Generalised or partial (focal).
- Pattern / types: clonic, tonic, tonic-clonic, atonic, myoclonic, startle, spasm, absence, others (contracting the thighs, pedaling, automatism) and status epilepticus.

Concepts & Glossary

Attack

- A violent act starting on with vigor.
- Used to describe epileptic & non-epileptic disorder.
- We say “attack of laughing” ; “Breath Holding Attack” ; “An attack of febrile convulsion” ; “Epileptic attack”.

Convulsion

- An intense, paroxysmal, involuntary muscular contraction with repetitive movements. Always implies movement.
- Is used for describing epileptic & non-epileptic motor act / shaking

Seizure

- A sudden attack or spasm.
- Mostly used to describe epileptic disorders.

Fit

- A seizure or convulsion especially one of epileptic nature.

A convulsive attack is always labeled as seizure. Contrary, not all seizures convulse and so, are not convulsions.

Duration of an Attack

The duration of the active part of the attack (its dominant pattern) from the start to its cessation.

Types of seizures

Tonic Type

- Usually generalized hypertonia (axial & of the 4 limbs).
- Fixed regard or up-rolling of eyes with apnea, erythrosis and cyanosis.
- Duration : mostly of few seconds.
- Rapid return to precedent activity with deep inspiration and/or cry.
- May be limited only to up-rolling of eyes. Usually one or repeated accesses.

Clonic Type

- A paroxysm (intense or not), of involuntary contraction or series of contractions of voluntary muscles, associated with LOC.
- Rhythmical jerky (shaking) movements of the 4 limbs (usually of upper limbs predominance).
- Sometimes of high frequency (very speedy), giving the impression of a tonic vibration (clonus). Some times of low velocity.
- Stopped by digressive manner up to sudden total abortion.
- May be associated with cyanosis, apnea, mouth frothy secretions and involuntary passage of urine.

Diff. Diagnosis:

- Shivering (from fear, high fever or cold) / can be aborted by gentle maneuvers.

Tonic-Clonic Type

- Consists of 2 phases. Starts as tonic and followed by clonic phase.
- This sequence is not always respected, giving other patterns: tonic-clonic-tonic, clonic-tonic-clonic.

Atonic Type “Drop attacks”

- All of sudden synchronous total loss of muscular tonus with loss of posture and LOC and dropping down if in vertical position.
- Sometimes it is associated with apnea, but not with movements.

NB. This is not an absence attack.

Diff. Diagnosis

- Faintness / syncope (progressive and not sudden).

Myoclonic Type

Myo from muscle and clony from movements

- Sudden startle (جفلة) of very brief duration (a fraction of a second) or generalized minor motor seizures which consist of abrupt, brief, jerky, one or two-sided.
- Usually take place in clusters of 2-3 over 1-2 seconds .
- Involve one muscle, a group of muscles, a segment of a limb or a whole limb.
- Usually involves the shoulder, but also the face mainly the outer part of the lips and the upper lids.

Types:

1. Physiologic myoclony:

- At the 1st phase of sleep and at awaking.
- Whole-mark / always during sleep.
- Anti-epileptic therapy / aggravation.

2. Pathological Myoclony:

- Audiogenic Myoclony / induced by sudden noise.
 - Tay-Sachs disease.
 - Krabbe disease .
- Cerebello-opso-myoclonic syndrome
 - During awake.
 - Ataxia.
 - Opsoclony (anarchic eye movements).

Infantile Spasms

A sudden contraction of all axial muscles with maintenance of this contraction over about 2 seconds. This is produced in repeated clusters, each is formed of a little number of consequent spasms. Between clusters there are periods of total muscular relaxation of about 5-20 seconds. The child may cry in between the attacks. There is no loss of consciousness.

Types:

1. Spasm in flexion

Synonym / Salam spasm

- Moro reflex-like.
-

2. Spasm in extension

- Incurving of the trunk.

- In both types there is contraction of both flexor and extensor muscles, but with a predominance of one.
- No prognostic difference between the 2 types.
- Asymmetry (head and eyes lateral deviation) is very indicative of cerebral lesion.

Diff. Diagnosis

- Manifestations of awaking .
- Gastro-esophageal reflux.
- Generalized Contraction due to abdominal pain.
- Moro reflex beyond 4 months.

Absence Type

Petit mal seizure

Are non-convulsive seizure consisting of brief (seconds) rupture of all contacts with the environment with interruption of all activity (freezing).

- Several episodes may occur in a day.
- Are associated with a highly specific 3-per-second spikes and waves on EEG .

Types:

1. Simple absence:

Not associated with any additional manifestations.

2. Complex absence:

Associated with other manifestations (clonic or myoclonic movements)

Status Epilepticus

It is defined as continuous seizure activity lasting longer than 30 minutes, or the occurrence of two or more seizures in quick succession without full return to usual level of consciousness during the inter-ictal intervals.

- Is an emergency that can result in neurological sequelae or death if treatment is delayed.

Main Question of Diagnosis in Case of a Seizure

1st Q:

Is it is seizure ? ... if not what is it? .. conversion reaction, tic, syncope, ...

2nd Q:

What type of seizure ? .. (clonic, tonic, tonico-clonic, atonic, absence,.....

3rd Q:

Is it epileptic or non-epileptic ? ... (FC, pseudoseizure, tetany,.....

4th Q:

If epileptic, which one ? (Generalized, partial, hemiconvulsion,...

Sensory System

Sensory system is a part of nervous system.

It consists of:

1. Sensory Receptors.
2. Neural pathways.
3. Parts of brain that processes this information.

1. Sensory Receptors

Specialized endings of afferent neurons (or separate cells that affect ends of afferent neurons) that collect stimuli from internal and external environment.

- Any energy form that activates a receptor is called **stimulus**.
- The process by which a stimulus is converted into an afferent electrical response is called **stimulus transduction**.
- Each receptor is specific to a certain type of stimulus, which is called its **adequate stimulus**. For example, photoreceptors can only detect light, while heat receptors can only detect heat, and pressure receptors can only detect pressure, etc.
- A single afferent neuron with all its receptor endings makes a **sensory unit**.
- Sensation is based on signal-transduction. The end-point is the same in all cases: processing of environmental information by membrane polarization resulting in the transmission of a nerve impulse.
- The information collected is called **sensory information**. It may or may not lead to conscious awareness. If it does, it can be called **sensation (senses)**.

Human sensory receptors:

Collect and transmit sensory impulses from:

- Skin
- Mucous membranes
- Muscles tendons
- Viscera

1. Chemoreceptor (Chemosensor)

A cell or group of cells that transduce a chemical signal into an action potential.

2 main classes are known:

a. Direct

(include taste buds in the gustatory system and carotid bodies that detect changes in pH inside the body).

b. Distance

(olfactory receptor neurons in the olfactory system).

2. Mechanoreceptor

A sensory receptor, at the peripheral (distal) endings of sensory neurons, that responds to mechanical pressure or distortion.

2 main types of cutaneous mechanoreceptors:

- a. **With small, accurate receptive fields**, found in areas needing accurate taction (e.g. the fingertips).
 - b. **With large, less accurate receptive fields**, found in areas needing less precise taction (i.e. the palm).
- Provide the senses of touch, pressure, vibration, proprioception and others.
 - Can also be separated into categories based on their rates of adaptivity. When a mechanoreceptor receives a stimulus it begins to fire impulses or action potentials at an elevated frequency (the stronger the stimulus, the higher the frequency). The cell, however, will soon “adapt” to a constant or static stimulus and the pulses will subside to a normal rate. Receptors that adapt quickly (i.e. quickly return to a normal pulse rate) are referred to as “**phasic / rapid**”. Those receptors that are slow to return to their normal firing rate are called “**tonic / slow**”.
- Phasic mechanoreceptors are useful in sensing such things as texture, vibrations, etc ; whereas tonic receptors are useful for temperature and proprioception among others.

3. Nociceptor

Is a sensory receptor that responds only after a high level of stimuli or a level enough to hurt the individual.

- When they are activated they can generate a reflex and pain.
- They respond to intense mechanical deformation, excessive heat, excessive lack of heat (e.g cold), electrical stimulation, many chemicals, and other forms of stimulation, the majority of which are damaging to the tissue surrounding the nociceptor.
- Pain and temperature sensation are extremely important to our survival. Therefore there are very complex connections which are even now poorly understood. Pain and temperature receptors are located in the dermis and epidermis of the skin and in the viscera. These receptors send synaptic projections to the dorsal root ganglion (DRG) which in turn projects via the dorsal horn of spinal cord, crosses to the lateral column of the spinal cord and ascends via the lateral spinothalamic tract to the thalamus. This thalamic projection accounts for much of the emotional response to chronic pain.
- A reflex arc exists in the spinal cord level, so that we withdraw our hand from a hot object even before we are aware that we have touched something hot.

4. Photoreceptor

Is a light-sensitive protein involved in the function of photoreceptor cells.

- Some examples are rhodopsin and photopsins in the human retina, phytochrome in plants, and bacteriorhodopsin in some bacteria.

5. Thermoreceptor

Is a sensory receptor that responds to heat and cold.

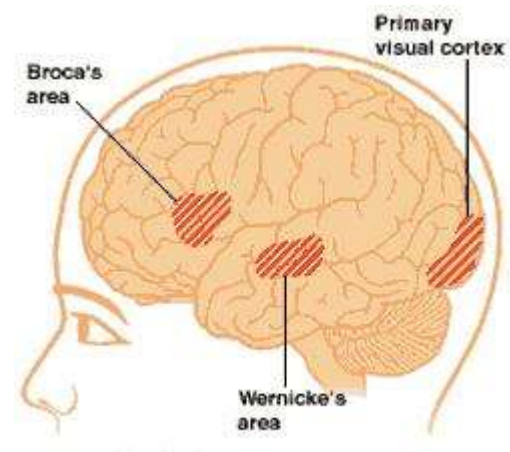
2. Neural Pathways

Conduct the sensory information to brain. Afferent neurons enter the CNS, diverge and synapse upon many inter-neurons.

- These afferent neurons are called **sensory or ascending pathways**. They are **specific ascending pathways** because they carry information of a single type of stimulus and from a single site.
- The ascending pathways reach the cerebral cortex on the **side opposite** to where their sensory receptors are located.

3. Parts of brain that processes this information

Specific ascending pathways that transmit information from somatic receptors and taste buds go to **somatosensory cortex** (parietal lobe), the ones from eyes go to **visual cortex** (occipital lobe), and the ones from ears go to **auditory cortex** (temporal lobe).



Sensory systems

1. Somatosensory
2. Olfactory
3. Visual
4. Hearing
5. Taste

As mentioned above, each is formed of specific sensory receptors with its neural pathways and a part of the brain that processes this specific information.

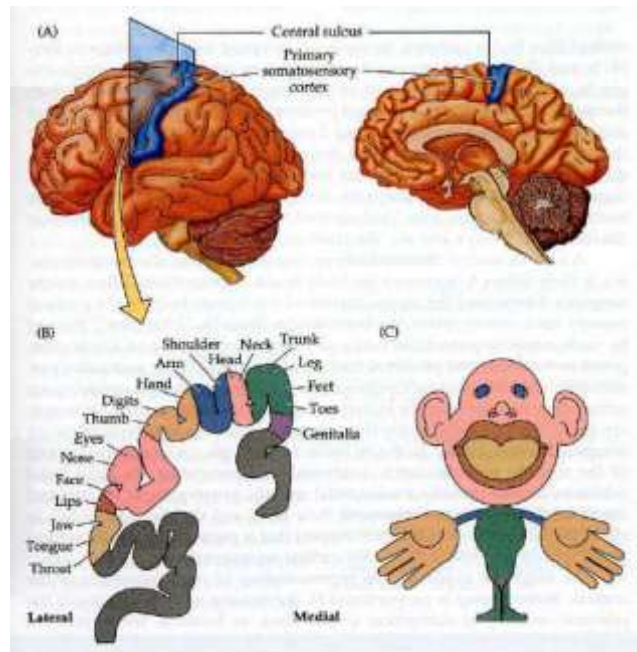
1. Somatosensory System

Is the sensory system of somatic sensation.

- Somatic sensation comes from various sensory receptors that trigger the experiences labeled as:

1. Touch
2. Pressure,
3. Temperature (warm or cold)
4. Pain (including itching الحكة and tickling دغدغه)
5. Proprioception (the sensations of muscle movement and joint position including posture, movement, facial expression).

- The somatosensory area in the human cortex is located in the postcentral gyrus. Areas of this part of the human brain map to certain areas of the body, dependant on the amount or importance of somatosensory input from that area. For example, there is a large area of cortex devoted to sensation in the hands, while the back has a much smaller area.



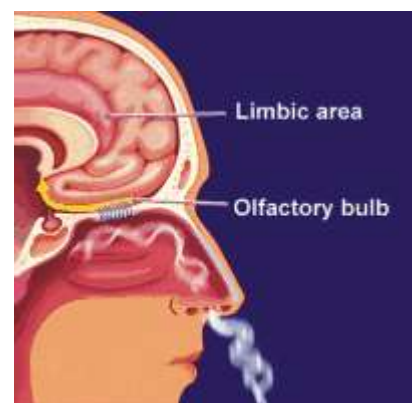
The **somatosenses** include:

1. Cutaneous (skin sensation).
2. Kinesthesia (movement sensation).
3. Organic sensation (sensory information from the organs, such as stomach aches).

2. Olfactory System

Thousands of odorants are smelled.

- Olfaction, the sense of smell, is the detection of chemicals dissolved in air.
- In vertebrates smells are sensed by the olfactory epithelium located in the nose and processed by the olfactory system.



3. Visual System

Is perhaps the best understood of the senses.

Two classes of photoreceptor cells exist in the retina: **rods** and **cones**.

The **rods** are more numerous, some 120 million, and are more sensitive than the cones. They are not sensitive to color.

The **cones** are about 6 to 7 million. They respond to bright lights and colors. Are much more concentrated in the central yellow spot known as the **macula**.

When light is absorbed, this triggers a sequence of reactions which lead to a nerve impulse. This is transmitted to the brain via the optic nerves.

4. Auditory System



- The immediate receptors for hearing are found in the hair cells of the cochleae. The mechanical motion of the cilia is converted into current flow and then into a nerve impulse.

3. Taste System

We can detect only five tastes:

- Bitter (مر),
- Sweet (حلو),
- Salt (مالح),
- Sour (حامض),
- Umami (taste of glutamate).



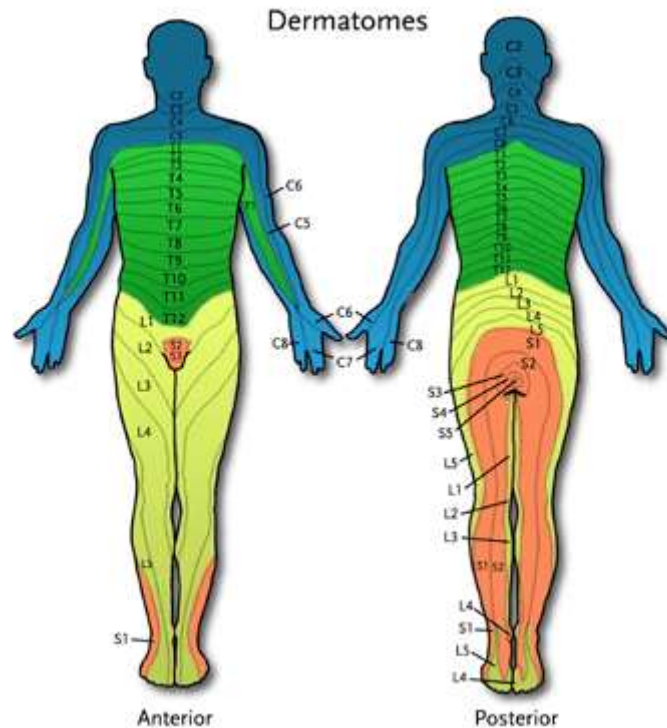
General Sensation

- Proprioception, fine touch and vibration is also a relatively straightforward pathway. Receptors are located in muscles, joints and tendons and project to the dorsal root ganglion (DRG). Fibers then ascend via the ipsilateral dorsal white column. The lumbosacral fasciculus ascends to nucleus gracilis and the thoracocervical fasciculus ascends to nucleus cuneatus. Both of these nuclei are located in the lower medulla. Fibers then cross as the medial lemniscus and finally ascend to the ventral posterior lateral (VPL) nucleus of the thalamus.
- General sensation from the face is mediated by the trigeminal nerve (cranial nerve V). It has three branches, The Ophthalmic (V1), Maxillary (V2) and Mandibular (V3) which project to the trigeminal ganglion in the pons and the trigeminal nucleus in the medulla. Fibers cross and ascend to the ventral posterior medial (VPM) nucleus in the thalamus.
- Fibers for pressure and crude touch synapse in the pons and also ascend to the VPM.

Dermatomes

The band of skin innervated by the sensory root of a single spinal nerve.

- Mapped / The map has been produced as accurately as possible, taking into account the slight variations and overlaps in sensory sectors
- Dermatomes overlap each other.



Clinical manifestations

1. Paraesthesia
pins and needles sensation in the hands and feet (إحساس بوخز خفيف).
2. Dysesthesia (pain).
3. Sensory loss (anesthesia and to a less degree hypoesthesia).

Examination of the Sensory System

The sensory exam includes testing for:

1. Pain sensation (pin prick)
2. Light touch sensation (brush)
3. Position sense
4. Stereognosis
5. Graphesthesia
6. Extinction

Sensory Testing / General Rules

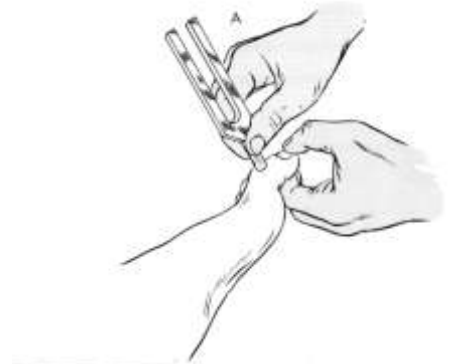
- Explain each test before you do it.
- Unless otherwise specified, the patient's eyes should be closed during the actual testing.
- Compare symmetrical areas on both sides of the body.
- Compare distal and proximal areas of the extremities.
- When you detect an area of sensory loss, map out its boundaries in detail.

Vibration



Use a low pitched tuning fork (128Hz).

- Test with a non-vibrating tuning fork first to ensure that the patient is responding to the correct stimulus.
- Place the stem of the fork over the distal interphalangeal joint of the patient's index fingers and big toes.
- Ask the patient to tell you if they feel the



vibration.

- If vibration sense is impaired proceed proximally: Wrists, elbows, medial malleoli, patellas, anterior superior iliac spines, spinous processes, clavicles

(may be skipped unless an abnormality is suspected).

Subjective Light Touch

Is a quick survey for "strange" or asymmetrical sensations only.



- Use your fingers or a fine wisp of cotton to touch the skin lightly on both sides simultaneously.
- Test several areas on both the upper and lower extremities.
- Ask the patient to tell you if there is difference from side to side or other "strange" sensations and whenever a touch is felt.

Pain & Light Touch Sensation



- This is an initial evaluation of the sensory system.
- Patient lying supine with eyes closed.
- Instruct the patient to say "sharp" or "dull" when they feel the respective object.
- Show the patient each object and allow them to touch the needle and brush prior to beginning to alleviate any fear of being hurt during the examination.
- With the patient's eyes closed, alternate touching the patient with the needle and the brush at intervals of roughly 5 seconds.
- Begin rostrally and work towards the feet.
- Make certain to instruct the patient to tell the physician if they notice a difference in the strength of sensation on each side of their body.
- Alternating between pinprick and light touch, touch the patient in the following 13 sites.
- Touch one body part followed by the corresponding body part on the other side (e.g., the Rt. shoulder then the Lt. one) with the same instrument. This allows the patient to compare the sensations and note asymmetry.

1	Posterior aspect of the shoulders	C4
2	Inner & outer aspect of the upper arms	C5-C6-T1
3	Medial aspect of the lower arms	T1
4	Tip of the thumb	C6
5	Tip of the middle finger	C7
6	Tip of the small finger	C8
7	Thorax, nipple level	T5
8	Thorax, umbilical level	T10
9	Upper part of the thigh	L2
10	Lower-medial part of the upper leg	L3
11	Medial lower leg	L4
12	Lateral lower leg	L5
13	Little finger & sole of foot	S1

If there is a sensory loss present, test vibration sensation and temperature sensation with the tuning fork. Also concentrate the sensory exam in the area of deficiency.

Position Sense

proprioception

- Grasp the patient's big toe and hold it away from the other toes to avoid friction.



- Show the patient "up" and "down."
- With the patient's eyes closed ask him to identify the direction you move the toe.
- If position sense is impaired move proximally to test the ankle joint.
- Test the fingers in a similar fashion.
- If indicated move proximally to the metacarpophalangeal joints, wrists, and elbows.

- Repeat on the opposite foot and

compare.

- Make certain to hold the toe on its sides, because holding the top or bottom provides the patient with pressure cues which make this test invalid.

- Fine touch, position sense (proprioception) and vibration sense are conducted together in the dorsal column system.
- Rough touch, temperature and pain sensation are conducted via the spinothalamic tract.
- Loss of one modality in a conduction system is often associated with the loss of the other modalities conducted by the same tract in the affected area.

Romberg's Test

Is a neurological test of balance

It is positive in patients with sensory ataxia.

The test is performed in two stages.

- 1st the patient stands with feet together, eyes open and hands by the sides.
- 2nd the patient closes the eyes while the examiner observes for a full minute.

Elicit whether the patient falls when the eyes are closed.

It is advisable to stand ready to catch the falling patient.

What that means "positive test" ?

- Romberg's test is **positive** if, **and only if**, the following 2 **conditions** are both met:
 - The patient **can stand with the eyes open.**
 - &
 - The patient **falls when the eyes are closed.**
- Romberg's test is **negative** if either of the following conditions are met:
 - The patient **falls when the eyes are open.**
 - or
 - The patient **sways يتأرجح** but **does not fall** when the **eyes are closed.**



The principle

Maintaining balance while standing in the stationary position relies on:



1. Intact sensory pathways:
 - a. Joint position sense / proprioception, carried in the dorsal columns of the spinal cord.
 - b. Vision.
2. Intact vestibular system.
2. Sensorimotor integration centers (cerebellum).
3. Motor pathways (corticospinal / pyramidal tract).

Interpretation of results:

- The **1st stage** of the test (standing with the eyes open), demonstrates that at least 1 of the 2 sensory pathways is intact, and that sensorimotor integration, vestibular system and the motor pathway are intact.
- In the **2nd stage**, the visual pathway is removed by closing the eyes:
 - a. If the proprioceptive pathway is intact, balance will be maintained, but
 - b. If proprioception is defective, both of the sensory inputs will be absent and the patient will sway then fall.

Remarks:

- Romberg's test is **not a test of cerebellar function**, as it is commonly misconsidered. Patients with cerebellar ataxia will generally be unable to balance even with the eyes open: therefore the test cannot proceed beyond the 1st step and no patient with cerebellar ataxia can correctly be described as Romberg's positive. Rather, Romberg's test is a test of the proprioception receptors and pathways function.

- Romberg's test is positive in conditions causing sensory ataxia such as:
 - a. Conditions affecting the sensory nerves (sensory peripheral neuropathies, such as chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)).
 - b. Conditions affecting the dorsal columns of the spinal cord such as tabes dorsalis.

Dermatomal Testing

- If vibration, position sense, and subjective light touch are normal in the fingers and toes you may assume the rest of this exam will be normal.

Pain

Use a suitable sharp object to test "sharp" or "dull" sensation (break a wooden cotton swab to create a sharp end. The cotton end can be used for a dull stimulus. Do not use non-disposable instruments such as those found in certain reflex hammers. Do not use very sharp items such as hypodermic needles.

Temperature

Often omitted if pain sensation is normal.

Use a heated or cooled object and ask the patient to identify "hot" or "cold."

Graphesthesia

- With the blunt end of a pen or pencil, draw a large number in the patient's palm.
 - Ask the patient to close their eyes.
 - Ask the patient to identify the number or letter you will write with the back of a pen on their palm.
 - Repeat on the other hand with a different letter or number.



Stereognosis

A higher cerebral associative cortical function reflecting the ability to determine what an object is, just by using the modality of touch.

- Ask the patient to close their eyes and feel an unknown object (coin or pen) placed into the hand.
- Repeat this with the other hand using a different object.

Astereognosis refers to the inability to recognize objects placed in the hand. Without a corresponding dorsal column system lesion, these abnormalities suggest a lesion in the sensory cortex of the parietal lobe.

- It is used as an alternative to graphesthesia.



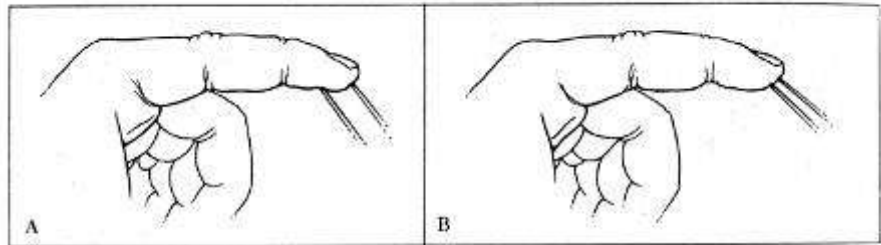
Two Point Discrimination

- Use in situations where more quantitative data are needed, such as following the progression of a cortical lesion.

- Use an opened paper clip to touch the patient's finger pads in two places simultaneously.

- Alternate irregularly with one point touch.

- Ask the patient to identify "one" or "two."



Two-point discrimination. A. The patient perceives the touch of two wooden sticks at 33 mm apart as two distinct stimuli. B. When the sticks are less than 3 mm apart, the touch is felt as though one stick were being used.

- Find the minimal distance at which the patient can discriminate.

Since the above tests (two point discrimination, stereognosis, graphesthesia) are dependent on touch and position sense, they cannot be performed when the tests for these senses are clearly abnormal.

Extinction



- Have the patient sit on the edge of the examining table.
- Close his eyes.
- Touch the patient on the trunk or legs in one place and then tell the patient to open their eyes and point to the location where they noted sensation.
- Repeat this maneuver a 2nd time, touching the patient in 2 places on opposite sides of their body, simultaneously.
- Ask the patient to point to where they felt sensation. Normally they will point to both areas. If not, extinction is present.

With lesions of the sensory cortex in the parietal lobe, the patient may only report feeling one finger touch their body, when in fact they were touched twice on opposite sides of their body, simultaneously. With extinction, the stimulus not felt is on the side opposite of the damaged cortex. The sensation not felt is considered "extinguished".